


## **Periodontal Manifestations of Local and Systemic Diseases**

Colour Atlas and Text

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George Laskaris · Crispian Scully

# Periodontal Manifestations of Local and Systemic Diseases

Colour Atlas and Text

With a Contribution by Dimitris N. Tatakis

Foreword by Prof. Jan Lindhe

With 392 Figures in Colour  
and Appendix with 8 Tables



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### Dedication to Christos Laskaris

Christos Laskaris was the son of Professor George Laskaris and his wife Vivi. Christos was a final year student in the School of Dentistry, University of Belgrade. He was a bright young 24 year old man with an engaging personality, full of energy and vitality, always polite and friendly, always having a good word to say for everybody, always smiling and willing to help. We had all hoped he would succeed his father as an outstanding stomatologist.

It was a tremendous misfortune to lose Christos so unexpectedly, and so unjustly, from a sudden illness.

This book is dedicated to his memory.

Professor Crispian Scully

This book is also dedicated to our families  
for their constant support of our efforts.

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## Foreword

I am delighted to see the appearance of the first comprehensive atlas recognizing the important relationship between periodontal and systemic health and disease. The atlas is not intended to cover the new areas of interest in relation to periodontal health and cardiac and other disease, but rather to demonstrate the systemic diseases that manifest to the periodontist and hygienist. The atlas thus covers a vast range of systemic disorders that may present with gingival or periodontal lesions and highlights the importance of the periodontal and oral medicine teams in diagnosing and managing patients with sometimes complex medical problems.

The authors, Professor George Laskaris and Professor Crispian Scully, are both active clinicians, teachers and researchers in oral medicine, with wide experience and special interest in the oral medicine/periodontal interface and both have published widely in their fields.

I welcome this contribution to the advancement of care for the patient with periodontal disease.

Jan Lindhe  
Professor Emeritus of Periodontology  
University of Gothenburg, Sweden

## Preface

The periodontium is a part of the oral tissues of great interest to dentists and particularly to the periodontal team. The importance has now been recognized to extend beyond local disorders to a wide range of conditions that may affect periodontal health.

The most recent classification proposed by the International Workshop for the Classification of Periodontal Diseases and Conditions in 1999 is testimony to an increasing scientific interest of the international dental community in periodontal pathology, and the wide range of local and systemic disease affecting the periodontium.

This atlas aims to illustrate and evaluate diseases targeting the gingivae and periodontium from the viewpoint of oral medicine and to increase further the awareness and interest of dentists, periodontologists and hygienists regarding the vast spectrum of diseases that can affect the gingivae and periodontium. It is also hoped that it will help open new scientific and professional avenues by demonstrating the important role of these professional groups in the art and science of medicine. The ultimate beneficiaries should be our patients – not least since several of the diseases may be life-threatening and require special expertise in early diagnosis and treatment.

Due to the greater variety of diseases affecting the gingivae and periodontium than was recognized in the above-mentioned workshop, we encountered a number of difficulties in adhering fully to that classification and thus elected to adopt a classification based on three main groups of pathologic entities: local diseases, manifestations of systemic disorders in the gingivae and periodontium, and cysts, tumours and neoplasms.

For each entity we present the clinical manifestations, frequency of involvement of the gingivae and other structures in the mouth and elsewhere, differential diagnosis, laboratory work-up, and principles of treatment. The illustrations are arranged so that for every disease there is one or more illustration demonstrating representative lesions of the gingivae. In addition, for the systemic diseases, we have often included photographs of lesions of other areas of the oral mucosa, extra-oral mucosal surfaces, or the skin, in order to better emphasize the spectrum of clinical manifestations.

We are certain that, with more comprehensive biological and medical education, dentistry will open new horizons in health services within the next few years. This was the motivation for the publication of the present work, and it is our hope that the atlas will help in the realization of this goal.

George Laskaris, Athens, 2002

Crispian Scully, London, 2002

**Acknowledgements**

We thank our patients and nurses who have taught us so much over the years, and continue to do so, and all those students and colleagues with whom we have worked and interacted, who may have shared the clinical care of some patients, and/or may have knowingly or otherwise contributed ideas or content. In this respect we thank especially Professors Oslei Paes de Almeida (Brazil), Jose-Vicente Sebastian-Bagan (Spain), Johann Beck-Managetta (Austria), Roman Carlos (Guatemala), Marco Carrozzo (Italy), Roderick Cawson (UK), Pedro Diz Dios (Spain), Dore Eisen (USA), Joel Epstein (Canada), Eleni Gagari (USA), Sergio Gandolfo (Italy), Andreas Katsabas (Greece), Christos Kittas (Greece), Jens Pindborg (Denmark; deceased), Stephen Porter (UK), Peter Reichart (Germany), Pierre-Luigi Sapelli (Italy), Sol (Bud) Silverman (USA), John Stratigos (Greece), Isaac Van der Waal (Netherlands) and Antony Vareltzidis (Greece). We are grateful to Dimitris Tatakis for contributing the chapter on Normal Periodontium, and to colleagues who contributed clinical illustrations, including Drs A. Parashis, N. Papadogeorgakis, S. Ashan, M. Glick, M. Griffiths, S. Porter, T. Hodgson, O. Almeida, M. Spostos, and N. Kumar.



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## PART I

**The Normal Periodontium**

Dimitris N. Tatakis

This chapter will review the normal periodontium (anatomy and physiology), the etiology and pathogenesis of gingivitis and periodontitis, and the diagnosis of periodontal diseases.

### Anatomy

**Periodontal Tissues** The following tissues, in order of encounter from the vestibular or oral aspect, constitute the periodontium and support the teeth: gingiva, alveolar bone, periodontal ligament and cementum. Under ideal healthy conditions, the gingiva is the only clinically observable periodontal tissue and the tissue on which most of the pathological manifestations occur. For this reason, the emphasis in this introductory chapter will be on the gingiva.

### Clinical Features of the Gingiva

The gingiva, i. e. the keratinized mucosa that covers the teeth and the alveolar bone, is demarcated from the alveolar mucosa by the mucogingival junction (MGJ) (**Figs. 1–5**). In health, the gingiva should have a light or salmon pink colour, compared to a much darker or red colour for the alveolar mucosa (**Figs. 1, 2**). Depending on the extent of skin pigmentation of the individual, the gingiva may also display significant pigmentation (**Fig. 4**).

The gingiva is divided into free or marginal and attached gingiva (**Fig. 5**). The free gingiva constitutes the movable, most coronal portion of the tissue. The attached gingiva is the immovable portion that extends to the MGJ. In less than 50 % of gingiva examined, a shallow groove runs parallel to the gingival margin and delineates the free from the attached gingiva (**Fig. 5**). This gingival groove is located 1–2 mm apical to the margin and its formation appears to depend on the organization of the underlying connective tissue collagen fibres. The attached gingiva often exhibits stippling (orange peel-like appearance) of the surface, most evident on the facial aspect of maxillary anterior teeth (**Figs. 2, 4**). In periodontally healthy subjects, the buccolingual dimension (thickness) of facial gingiva ranges between 0.80 mm (mandibular canines) and 1.52 mm (mandibular 2nd molars). The apicocoronal dimension (width) of the attached gingiva ranges between 1 and 10 mm and varies by site. It is the narrowest on the lingual of mandibular incisors, the facial of mandibular canines and first bicuspid, and the facial of maxillary first bicuspid. It is widest on the facial of maxillary incisors and the lingual of mandibular molars.



**FIGURE 1**

Normal periodontium.  
The gingiva appears with a typical  
salmon pink colour

**FIGURE 2**

Normal periodontium.  
Note stippling of attached gingiva

**FIGURE 3**

Normal periodontium.  
The mucogingival junction is  
demonstrated by iodine staining  
of the alveolar mucosa  
(same case as Fig. 2)



FIGURE 4

Normal periodontium.  
Normal pigmentation in an individual with moderate skin pigmentation. Note stippling of attached gingiva



FIGURE 5

Normal periodontium.  
The free and attached gingiva and the gingival groove are evident, particularly on the incisors



### Histology of the Gingiva

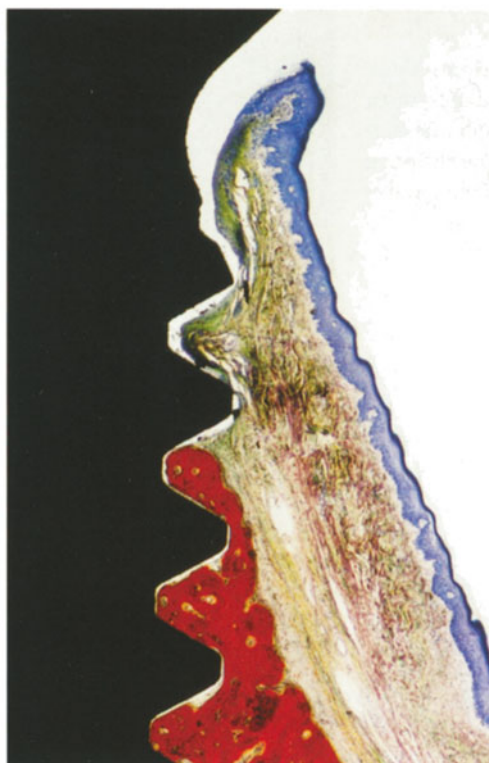
The gingiva consists of epithelium and connective tissue (**Fig. 6**). The gingival epithelium, according to location and histological characteristics, can be divided into three types: oral, sulcular and junctional epithelium.

The oral epithelium (OE) lines the surfaces facing the oral cavity. The OE is thick, stratified, orthokeratinized or parakeratinized, with elongated, narrow pegs (projections). The epithelium is separated from and linked to the underlying connective tissue by a basement membrane (basal lamina). The underlying lamina propria is dense and firmly attached to the periosteum without distinct submucosa.

The connective tissue of the gingiva is primarily composed of fibroblasts, collagen and elastin fibres, vasculature and neuronal processes. Other cells normally present include mesenchymal and immune system cells, e.g. monocytes, macrophages, neutrophils, lymphocytes and plasma cells. The collagen fibres of the gingival connective tissue are either randomly scattered in the extracellular ground substance or organized in characteristically oriented bundles. These bundles are named based on

FIGURE 6

Animal histology of gingiva around a titanium dental implant. The epithelium is stained *blue*, the connective tissue is *green*, and the alveolar bone is *red*



the location and direction of the fibres. Circular, dentogingival, dentoperiosteal, and transseptal fibres are the main groups described.

The sulcular epithelium is the non-keratinized epithelium with shallow pegs that lines the sulcus and faces the tooth. Its stratified structure resembles that of oral epithelium, except for the lack of complete keratinization. It continues into the junctional epithelium, which is the epithelial tissue attached to the tooth (enamel, cementum or dentine) and located at the bottom of the sulcus.

The junctional epithelium (JE), whose length is typically 1–3 mm, is a non-keratinized, stratified epithelium that is only one to two layers thick at its apical end and a few layers thick at its coronal end, i.e. at the base of the sulcus. The connective tissue underlying the JE is highly vascularized. Beneath the JE, inflammatory cell infiltrates can be observed even in clinically healthy gingiva.

The distance between the depth of the gingival sulcus, i.e. the most coronal limit of the junctional epithelium, and the alveolar crest has been termed biological width. It consists of the sum of the junctional epithelium (epithelial attachment) length and the length of the connective tissue attachment located apical to the junctional epithelium and coronal to the alveolar crest. The biological width reportedly ranges from 0.75 to 4.33 mm, while the epithelial attachment and the connective tissue attachment ranges are 0.32 to 3.27 mm and 0.29 to 1.84 mm, respectively. The mean values reported are 1.14 mm for the epithelial attachment and 0.77 mm for the connective tissue attachment. The biological width may be significantly greater in teeth treated for periodontitis.



FIGURE 7

Alveolar bone.  
The alveolar bone proper  
(socket wall) and its numerous  
openings are evident



**Alveolar Bone** The alveolar bone, which is clinically visible only in pathological conditions, has a structure similar to bone elsewhere in the body. The alveolar bone proper is the thin compact bone forming the walls of the tooth socket, and radiographically corresponds to the structure called lamina dura (Fig. 7). The alveolar bone proper is the bone that anchors the periodontal ligament fibres (Sharpey's fibres, see below). The wall of the alveolus shows numerous openings that allow passage of vessels and nerve fibres (Fig. 7).

Under ideal conditions, the crest of the interdental bone is located 1–2 mm apical to the cemento-enamel junction (CEJ) of the adjoining teeth. The shape and location of the crest depends on the width of the interdental space and the relative position of the CEJs.

The buccal and lingual compact bone plates of the alveolar process surround trabecular bone. The trabeculation pattern of this bone varies widely, and depends on functional conditions. Like all other bone, alveolar bone undergoes constant remodelling, which entails both bone formation and bone resorption. Systemic (metabolic) bone diseases may affect alveolar bone in a manner similar to their effect on long bones.

**Periodontal Ligament** The periodontal ligament (PDL) is a specialized loose connective tissue located between the alveolar bone and the tooth. PDL is the tissue that attaches the tooth (cementum) to the alveolar bone. It is a highly vascularized and innervated tissue with considerable adaptive capacity in response to mechanical stimulation. This property can probably be attributed to the very high turnover rate of the PDL connective tissue. The PDL contains precursor cells that give rise to bone-forming (osteoblasts) and cementum-forming (cementoblasts) cells. In addition to the above connective tissue cellular elements, PDL contains epithelial cells, the epithelial rests of Malassez. These remnants of the enamel organ (Hertwig's epithelial root sheath) are thought to contribute to cyst formation. Recent studies suggest that the epithelial rests may have functions related to the development of PDL innervation.

The principal collagen fibres of the PDL are organized in characteristically oriented bundles: the dentoalveolar crest group, the horizontal group, the oblique group, the apical group and the interradicular group.

The principal PDL fibres that are embedded in the alveolar bone and in cementum are called Sharpey's fibres.

The space between tooth and alveolar bone that is occupied by the PDL (PDL space) can vary, on average, from 0.08 mm for impacted teeth to 0.2 mm for teeth under function. There is wide variation, however, among individuals and teeth. The PDL space and changes in PDL space width, either in response to mechanical forces or as a consequence of diseases that significantly affect collagen turnover (e.g. scleroderma), can be visualized radiographically.

**Cementum** Although it is part of the tooth structure, cementum is also considered a tissue of the periodontium, as it helps attach the tooth to the gingiva and the alveolar bone. Cementum (and/or dentine) is clinically exposed when gingival recession is present (**Fig. 8**).

Cementum is found in two forms: acellular and cellular. Acellular cementum, as the name implies, does not contain embedded cementoblasts (cementocytes). It covers the cervical portion of the root and extends apically to a varying extent. The thickness of acellular cementum varies from a few microns near the cervix up to 200  $\mu$ m towards the apex. Cellular cementum contains embedded cementocytes and thus resembles bone in structure. Cellular cementum thickness increases with age and may reach several millimetres. Although it shares structural characteristics with bone, unlike bone, cementum lacks vasculature, exhibits continuous apposition throughout life and does not normally undergo remodelling. Pathological resorption may occur under several different conditions, including excessive mechanical stimulation. Such resorption may be followed by repair, i.e. deposition of cellular cementum.

Cementum may also be visualized in radiographs, especially when significantly increased in thickness (hypercementosis).

**Vasculature and Innervation** As alluded to previously, several periodontal tissues exhibit a high degree of vascularization. The gingiva receives its blood supply from suprapariosteal vessels as well as vessels emerging from the alveolar bone. The gingival blood vessels are typically arterioles, small veins and capillaries. Underlying the sulcular and junctional epithelium is a rich plexus of arterioles and venules. The PDL is the other highly vascularized periodontal tissue, with vessels originating from the alveolar bone and the dental (periapical) vasculature. The PDL vessels predominantly run parallel to the long axis of the tooth and are located closer to the bone surface than the cementum.

The gingiva is supplied by nerve fibres stemming from the trigeminal nerve (second and third division). These fibres reach the gingiva either from the suprapariosteal area (like the vessels) or through the coronal portion of the PDL.

The PDL is richly innervated by fibres originating either in the trigeminal ganglion or the mesencephalic trigeminal nucleus. These demonstrate differential distribution along the root. Ruffini-like nerve endings provide the PDL with mechanoreceptor function. Besides Ruffini-like endings, lamellated corpuscles and free nerve endings have been described in the PDL.

FIGURE 8

Clinically evident cementum and/or dentine because of recession in the canine and premolar area



### Biochemistry and Physiology

In animal studies, the quantitative composition of the gingiva has been reported to be 39 % epithelium, 5 % infiltrated connective tissue (ICT) and 56 % non-infiltrated connective tissue (NCT). ICT is present even in clinically healthy gingiva; the inflammatory infiltrate is composed of polymorphonuclear leukocytes (neutrophils), monocytes and macrophages, lymphocytes and plasma cells.

Water accounts for 66 % of the gingival tissue, while collagen fibres occupy 70 % of the connective tissue volume of NCT and only 18 % of the volume of ICT, indicative of the significant loss of collagen associated with inflammation of the gingiva. This loss clinically translates to the “transformation” of a very firm and dense tissue (healthy gingiva) to a friable and oedematous one.

Although collagen type I is the most abundant collagen type present in the connective tissue, with collagen type III being the second most prevalent type, other collagen types (e.g. IV, V, VI and XII) have also been found in the periodontium. Collagen types I and III constitute the bulk of the gingival fibres, while type V collagen filaments run parallel to the main fibres. In PDL, type I collagen is the main constituent of the principal fibres, although collagen type III, V, VI and XII are also found. Type I and III collagens are the principal component of cementum and alveolar bone organic matrix.

Besides collagen, glycosaminoglycans (e.g. dermatan sulphate, heparan sulphate, chondroitin sulphate, hyaluronate) and other extracellular matrix proteins (e.g. fibronectin, elastin, tenascin) contribute to the formation of the organic matrix. The organic matrix of the mineralized tissues contains several non-fibrous proteins such as osteopontin and bone sialoprotein-II. The organic matrix of alveolar bone and cementum also contains growth factors (biologically active protein molecules) which may be significant for tissue repair and regeneration. Examples of such polypeptides include platelet-derived growth factors (PDGFs), fibroblast growth factors (FGFs), transforming growth factors (TGFs), and bone morphogenetic proteins (BMPs).

The gingiva has a high turnover rate. It has been estimated that the turnover time for gingival epithelium is 10–12 days, while the reported



half-life of mature collagen is approximately 10 days in the lamina propria of the gingiva.

When subclinical inflammation is present in the gingiva, a fluid appears in the gingival crevice. This gingival crevicular fluid (GCF) is an exudate, with a composition that represents roughly a 30% dilution of serum. Besides serum proteins, GCF contains proteins produced locally in the gingiva, e.g. immunoglobulins synthesized and released by locally residing plasma cells.

### Microbiology and the Role of Microorganisms in the Pathogenesis of Periodontal Disease

Colonization of the oral cavity with microorganisms occurs during birth or soon thereafter, through contact. Establishment of common plaque species such as *Streptococcus sanguis* is evident after tooth eruption. Studies have shown that pathogenic plaque bacteria, whether cariogenic or periodontopathogenic, are transmitted from parents to children.

The classic experimental gingivitis studies of L  e and coworkers in the 1960s provided proof that dental plaque is the etiological factor for gingivitis, a reversible condition. Subsequently, human epidemiological studies and animal experiments showed that plaque accumulation is a factor that leads to the development of periodontitis, i.e. attachment loss, which is an irreversible condition.

The development of bacterial plaque, as it accumulates and matures, progresses from a supragingival collection of aerobic or facultative, Gram<sup>+</sup> cocci and non-motile rods, to a subgingival aggregation of anaerobic, Gram<sup>-</sup>, motile rods and other morphotypes. The organization of dental plaque in a biofilm, defined as a “matrix-enclosed bacterial populations adherent to each other and/or to surfaces or interfaces”, has special implications for the survival and management of the oral flora, especially in relation to the pathogenesis and treatment of periodontitis.

The normal flora, or the flora that is compatible with periodontal health, comprises primarily Gram<sup>+</sup> facultative microorganisms. This flora includes *S. sanguis*, *S. mitis*, *Actinomyces naeslundii*, *A. viscosus*, and *Veillonella parvula*, among other species.

The development of gingivitis is associated with an increase in Gram<sup>-</sup> species. Species associated with gingivitis include: *Fusobacterium nucleatum*, *Treponema denticola*, *Prevotella intermedia*, *Campylobacter rectus*, *Actinomyces*, and *Veillonella*.

In periodontitis, the subgingival flora is characterized by a significant shift to Gram<sup>-</sup> anaerobic species, such as *Porphyromonas gingivalis*, *Bacteroides forsythus*, *Actinobacillus actinomycetemcomitans*, *Campylobacter rectus*, *P. intermedia*, *Eikenella corrodens*, and *Treponema* species. A few of these bacteria have been more strongly implicated in certain forms of periodontitis, e.g. *A. actinomycetemcomitans* in localized aggressive periodontitis (previously classified as localized juvenile periodontitis, see “Diagnosis of Periodontal Diseases”). Certain species exhibit strong associations with each other, such as *P. gingivalis*, *B. forsythus*, and *T. denticola*; these bacteria are often found together and have been strongly associated with severe periodontitis.

Advances in PCR (polymerase chain reaction) cloning of subgingival bacteria have resulted in the identification of both cultivable and non-cultivable bacteria from clinical specimens. PCR results show a greater complexity of subgingival microbiota than previously realized. It seems likely that non-culture-based microbial analysis will identify new periodontal



pathogens. Recent studies have also raised the possibility that certain viruses may contribute to the pathogenic process of some forms of periodontitis.

## Pathology – Immunological Mechanisms

### Pathology and Clinical Correlates

Quantitative analysis of human gingival biopsies from experimental gingivitis and periodontitis studies has led to the description of four histopathological stages in the development of the disease.

The initial lesion develops within 4 days of the first exposure to plaque and is characterized by acute inflammation, vasculitis, migration of polymorphonuclear neutrophils (PMNs), alteration in the coronal aspect of the junctional epithelium (JE) and loss of collagen around blood vessels. The clinical feature of this stage is increased gingival crevicular fluid flow.

The early lesion has, in addition to the features of the initial lesion, the following characteristics: accumulation of lymphocytes, altered (damaged) fibroblasts, further loss of interstitial collagen and proliferative changes in the basal cell layer of JE. The collagen loss in the infiltrated area may be as high as 70 %. The predominance of lymphocytes is the distinguishing feature of this stage, which develops within 7 days of the beginning of plaque accumulation and is clinically evident as gingivitis.

As time progresses, the early lesion evolves into the established lesion, which clinically corresponds to chronic gingivitis. This stage is characterized by continued presence of inflammation, appearance of plasma cells and immunoglobulins in the tissue, connective tissue loss, and severe changes in the junctional epithelium. The duration of the established lesion is indeterminate; chronic gingivitis may persist for a long time as such, without ever progressing to periodontitis.

The advanced lesion corresponds to periodontitis clinically and has, in addition to the features of the established lesion, the following characteristics: alveolar bone loss, pocket formation (attachment loss), distant fibrosis and altered plasma cells.

Increased flow of gingival crevicular fluid (GCF) is the first clinical sign to appear during development of gingivitis. GCF flow correlates well with the degree of histological inflammation. Another clinical sign of gingival inflammation is bleeding on probing. Bleeding sites, when compared to non-bleeding sites, exhibit significantly higher levels of inflammation.

### Immunological Mechanisms – Immunopathogenesis

The epithelium of the gingiva forms the first natural barrier of the periodontium against microorganisms and foreign substances. The pathogenic potential of certain bacteria implicated in periodontitis, e.g. *P. gingivalis*, appears to be related to their ability to invade epithelial cells.

Polymorphonuclear leukocytes (PMNs or neutrophils) are an essential component of the innate or non-specific immune response against bacteria. Systemic conditions associated with periodontal involvement, such as localized aggressive periodontitis, diabetes, and cyclic neutropenia among others, are characterized by qualitative or quantitative PMN defects. Hereditary PMN disorders result in severe susceptibility to bacterial infections, in general, and periodontal infections, in particular.

Although PMNs are critical for periodontal health, evidence also suggests that these cells may be responsible, at least in part, for some of the tissue destruction encountered in periodontal diseases; in GCF or diseased gingiva from periodontitis patients, the collagenase present is pre-

dominantly of PMN origin. This has led to the pursuit of pharmacological anti-collagenase interventions.

Besides PMNs, macrophages and natural killer (NK) cells are also cellular components of the innate immune responses. Upon their migration from the vessel into the tissues, monocytes differentiate into macrophages, which contribute to pathogen elimination by phagocytosis. Hereditary defects in monocyte function, such as leukocyte adhesion deficiency syndromes, may lead to severe periodontal involvement (e.g. generalized aggressive periodontitis in children).

The adaptive or specific immune response relies on lymphocyte function. The gingiva contains significant numbers of the various lymphocyte classes and subclasses, distributed in the lamina propria and the infiltrated gingival connective tissue. Lymphocytes are categorized according to their function into B lymphocytes (B cells) and T lymphocytes (T cells). B cells, when appropriately stimulated, differentiate into plasma cells. The presence of high numbers of plasma cells in the infiltrated connective tissue is considered characteristic of long-standing periodontal pathology. T cells are classified as cytotoxic T cells and helper T cells. T helper cells have been classified into two major phenotypes, Th1 and Th2 cells, based on their functional properties, including the profile of cytokines produced. Some of the cytokines (see below) lead to pronounced inflammatory reactions, while others suppress inflammatory processes. The overall characteristics of the gingival immune (inflammatory) response to plaque bacteria appear to depend on the balance between Th1 and Th2 responses. Genetic predisposition to the development of responses with an exacerbated inflammatory component may lead to greater tissue destruction, i.e. severe periodontitis.

Severe immunodeficiencies, such as the congenital lack of both T and B cells, lead to lethal infections. Acquired immunodeficiencies, such as the severe depletion of T cells following HIV infection (AIDS), are also characterized by high susceptibility to infection, including the development of necrotizing ulcerative periodontitis.

Cytokines, protein factors produced by cells that affect other cells, have emerged as critical components in the immune response to plaque and the ensuing periodontal tissue destruction. Experimental periodontitis studies indicate that interleukin-1 (IL-1) and tumour necrosis factor (TNF) play a major role in the development of local inflammatory infiltrates and the accompanying alveolar bone loss. Interleukin-8 (IL-8) and monocyte chemoattractant protein-1 (MCP-1) are two cytokines that direct recruitment of PMNs and monocytes, respectively, to specific tissue sites such as the junctional epithelium and the subepithelial connective tissue. It appears that pathogens, such as *P. gingivalis*, may have the ability to inhibit local IL-8 production, which results in a lack of PMN recruitment and persistence of the pathogen. A lack of interleukin-10 (IL-10), a cytokine with significant anti-inflammatory properties, leads to more severe periodontal bone loss, as indicated from animal studies. Therefore, the relative balance between pro-inflammatory (IL-1, TNF) and anti-inflammatory (IL-10) cytokines may determine the severity of inflammation and tissue destruction.

Lipid molecules with pro-inflammatory activities such as the arachidonic acid metabolites (prostaglandins, thromboxanes, and leukotrienes) have been shown to be involved in inflammatory processes that lead to significant tissue remodelling or tissue destruction. The processes in question may be physiological (e.g. orthodontic tooth movement) or pathological (e.g. gingivitis, periodontitis) in nature. Use of non-steroidal anti-inflammatory drugs (NSAIDs), whose mechanism of action includes inhibi-

tion of prostaglandin synthesis, prevents the development of experimental gingivitis and halts the tissue damage associated with periodontitis.

Collectively, these findings suggest that control of the various inflammatory processes can prevent/limit the tissue destruction of periodontitis and prevent/reverse the tissue changes observed in gingivitis.

## Diagnosis of Periodontal Diseases

### Periodontal Examination and Diagnosis

The periodontal examination should include a written medical and dental history, a patient interview and a clinical and radiographic examination. The patient should be given the opportunity to consent to the examination procedures by signing an appropriate informed consent.

#### Self-Reported Medical and Dental History, Patient Interview

**Self-Reported Medical History.** This textbook is testament to the fact that numerous systemic conditions have important manifestations in the periodontium, in general, and the gingiva, in particular. Therefore, the significance of a complete medical history cannot be overstressed. Use of a written questionnaire to assess medical/dental history is essential. It should include a review of systems, with an emphasis on diseases known to affect the periodontal tissues and on conditions that may interfere with or complicate treatment. It should also elicit information on pertinent social/environmental factors (e.g. use of tobacco products, occupational exposure to hazardous materials). A list of relevant medications should be included, with a check/circle option.

**Self-Reported Dental History.** Identification of previous periodontal involvement/treatment, oral hygiene practices, and other relevant dental conditions and treatment (e.g. tooth extractions, orthodontic treatment, removable partial dentures) that may impact periodontal health should be included.

**Patient Interview.** During the interview process, the patient's chief complaint should be established. Then medical and dental histories are reviewed. Details regarding the timing and reasons behind any significant findings in the medical (e.g. specific drug treatment) or dental (e.g. time and reason for previous tooth extractions) history should be elicited. The interview process should be structured in a manner that helps ascertain the patient's dental/periodontal IQ (intelligence quotient) and treatment expectations. At the end of the interview, the practitioner should have a clear view of the patient's relevant or contributory past medical and dental history, of the need for any further medical clearance or consultation, and possibly of the need for medical or dental specialist referral.

#### Clinical Examination

The clinical examination should include recording of vital signs (blood pressure, pulse) and examination of extraoral and intraoral tissues. The extraoral exam should include evaluations of the skin and appendages, of the lips, of the temporomandibular joints (TMJ) and assessment of possible lymph node involvement. The intraoral exam should include visual assessment of all structures. Where necessary, tactile assessment (palpation) should also be performed. If indicated by the patient's chief complaint/medical history or by clinical findings, additional vital signs (e.g. temperature and respiration rate) should be recorded.



The proper examination of the periodontium requires charting of the clinical parameters described below on appropriate form(s). The examination should start with visual assessment of the gingiva, alveolar mucosa and teeth (Figs. 1, 2). Deviations from normal colour and surface anatomy or structure are recorded. In some patients this may require the removal of loose debris, food particles, plaque accumulations or excessive supra-gingival calculus from certain areas. The presence of such material is recorded, as well as the presence of factors that lead to plaque retention. Such factors may be iatrogenic as in inadequate restorative treatments (e.g. amalgam overhangs, open crown margins), pathological as in carious lesions, and anatomical/developmental in nature such as deep radicular grooves, enamel projections, and enamel hypoplasia. Although visual inspection may provide abundant information, the proper detection of these factors requires detailed examination with a sharp explorer.

After the visual assessment, periodontal probing is performed. Typically, probing should be recorded in six sites per tooth (mid-buccal, distobuccal, distolingual, mid-lingual, mesiolingual and mesiobuccal). However, the entire circumference of the tooth should be probed for proper examination and data collection. Using a periodontal probe with appropriate length marks, the depth of the gingival sulcus is measured to the nearest mm (probing depth: PD). In periodontal health, the probing depth varies between 0.5 and 4.5 mm, depending on the site and the individual (Fig. 9). It should be pointed out that evidence from histological studies indicates that the clinical probing depth does not necessarily correspond to the histological sulcus (or pocket) depth. This is because the tip of the probe may easily penetrate through the sulcular or junctional epithelium, depending on the applied force, the size of the instrument, and the inflammatory status of the tissues.

From a practical standpoint, probing is what is usually performed and recorded. Should one need to have more detailed information, then clinical attachment level (CAL) measurements will be required. Concurrently with PD, the distance from the free gingival margin (GM) to the cemento-enamel junction (CEJ) is measured (GM-CEJ). This measure, in conjunction with the probing depth, allows determination of the clinical attachment level (CAL). The formula used is  $CAL = PD - (GM - CEJ)$ . In the event of marginal tissue recession, the GM-CEJ distance is given a negative value. In this manner, in sites with gingival recession, CAL equals the sum of PD and recession (GM-CEJ). The amount of CAL is used to determine the severity of periodontitis (e.g. slight, moderate and severe for CAL levels of 1–2, 3–4, and 5, respectively). During probing, sites exhibiting bleeding on probing (BOP) or presence of purulent exudate (pus) are also recorded.

Probing should be performed with the probe held parallel to the long axis of the tooth (Fig. 9). In the interproximal areas, the probe needs to be slightly angulated to reach the deepest site under the contact point. When calculus is present, the probe should be gently guided past the deposit to reach the bottom of the pocket. In sites with significant calculus deposits, probing may be difficult or impossible and result in inaccuracies. In addition, coexistent severe gingival inflammation may render probing unfeasible due to patient pain sensitivity. In such cases, debridement and/or anaesthesia may be necessary prior to periodontal probing and charting.

Furcation involvement and tooth mobility should be assessed and recorded. Furcation involvement is evaluated using a curved periodontal probe (Fig. 10). Furcations are usually classified as Class I, II or III, depending on degree of involvement, i.e. loss of attachment in the interradicular area in a horizontal direction. Tooth mobility is classified as 1, 2 or 3, depending on increasing severity. Typically, each tooth is held between two

FIGURE 9

Periodontal probing.  
The probe is held parallel to the  
long axis of the tooth

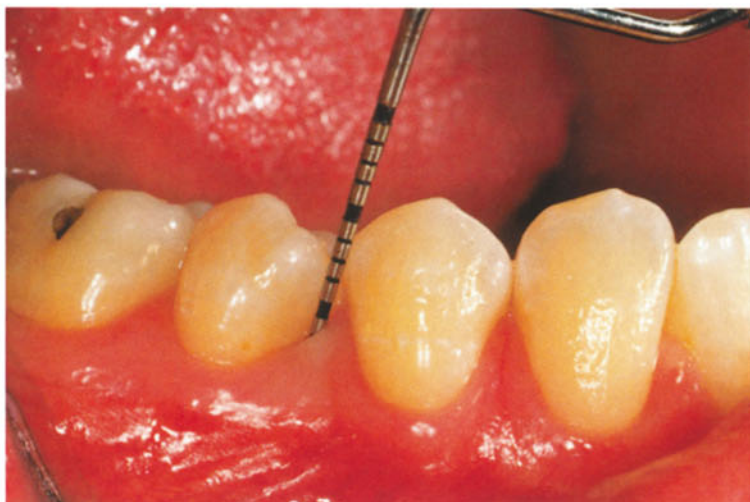


FIGURE 10

Furcation examination.  
The curved probe is used to detect  
furcation involvement



FIGURE 11

Determination of tooth mobility.  
The tooth is held between  
the two instruments





hard instruments and pushed in a labiolingual (and apical) direction to determine mobility (**Fig. 11**).

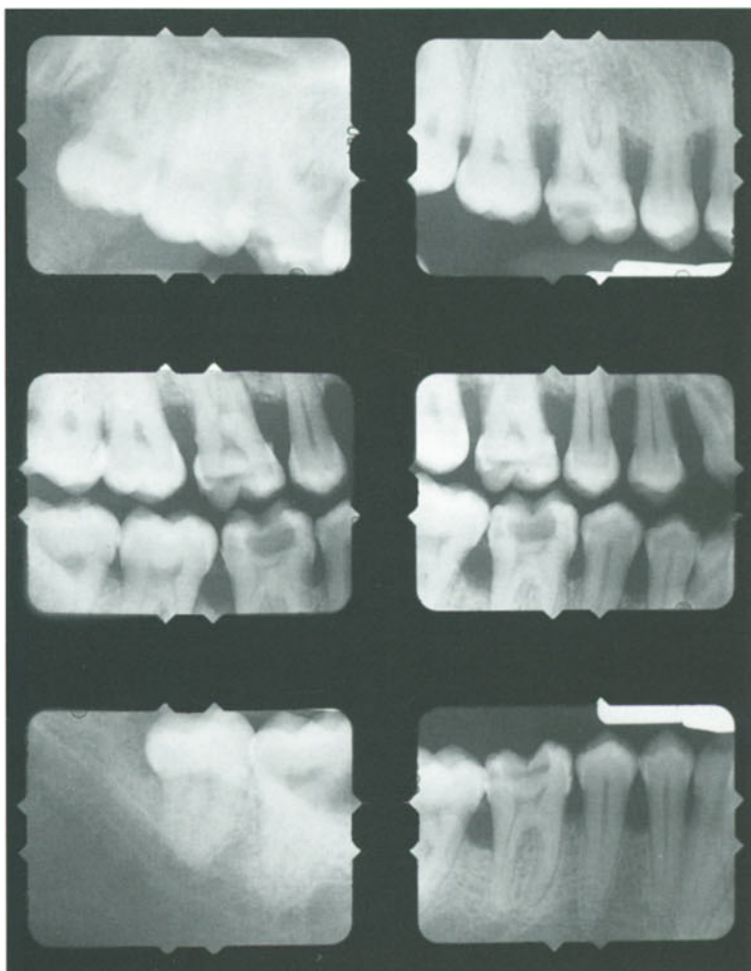
The radiographic examination (see “Radiographic Examination”) provides a two-dimensional depiction of the alveolar crest levels around the teeth. However, should the practitioner desire detailed information regarding the osseous anatomy, e.g. prior to surgical treatment planning, then bone sounding may be performed. Bone sounding, which requires anaesthesia, consists of probing through the gingiva (or mucosa) until resistance from the bone is met. Bone sounding allows the health care provider to identify the shape and dimensions of osseous defects clinically (e.g. intrabony defects) without reflecting a flap. It helps corroborate and expand on information provided by the radiographic examination and is useful in planning the surgical treatment of periodontal defects.

### Radiographic Examination

The radiographic evaluation is an integral part of the periodontal examination, as radiographic findings complement and corroborate clinical findings. The purpose of the periodontal radiographic examination is to determine the level of the alveolar bone crest and to identify changes in the hard (bone and teeth) and soft (PDL) periodontal tissues that cannot be visualized clinically. Radiographs also help identify calculus deposits, inadequate restorations (overhangs, etc.) and carious lesions on teeth.

**FIGURE 12**

Radiographic examination. Periapical and bitewing radiographs of right sextant. The patient, a 24-year-old, systemically healthy, non-smoker, female, exhibits moderate to severe bone loss. Calculus deposits are evident on the bitewing radiographs. The patient was diagnosed with generalized aggressive periodontitis



The best determination of alveolar bone levels is made from a full-mouth set of periapical and bitewing radiographs of diagnostic quality (Fig. 12). Ideally, vertical bitewing radiographs should be taken. In periodontal health, the alveolar crest level is located 1–2 mm apical to the CEJ; the interdental crest is also parallel to a line connecting the CEJ of two adjacent teeth. In cases of severe periodontitis, only vertical bitewing radiographs will allow depiction of the interdental alveolar crest levels, which may be located several millimetres apical to the CEJ (Fig. 12). Depending on the presence of specific lesions, additional types of radiographic evaluation, e.g. occlusal radiographs or CT scans, may be necessary.

### Diagnosis

At the end of the periodontal examination, the oral health care provider should reach a working (clinical) diagnosis based on the history, symptoms and signs present. He or she should ascertain the need for any laboratory tests or medical and dental specialist referral that may be necessary for definitive diagnosis and management of the case.

In 1999, the American Academy of Periodontology organized the International Workshop for a Classification of Periodontal Diseases and Conditions. At the conclusion of this workshop a new classification system of periodontal diseases was adopted (Table 1). In this new classification scheme, which is more comprehensive than previous ones, there are eight major categories, including gingival diseases, chronic periodontitis, aggressive periodontitis, periodontitis as a manifestation of systemic diseases, necrotizing periodontal diseases, abscesses of the periodontium, periodontitis associated with endodontic lesions, and developmental or acquired deformities and conditions. Of all the conditions listed, gingivitis associated with dental plaque and generalized chronic periodontitis are the two most common periodontal diseases.

TABLE 1

Classification of periodontal diseases  
(from Armitage 1999)

### I. Gingival Diseases

#### A. Dental plaque-induced gingival diseases

1. Gingivitis associated with dental plaque only
2. Gingival diseases modified by systemic factors
  - a. Associated with the endocrine system
    - 1) Puberty-associated gingivitis
    - 2) Menstrual cycle-associated gingivitis
    - 3) Pregnancy-associated
    - 4) Diabetes mellitus-associated gingivitis
  - b. Associated with blood dyscrasias
    - 1) Leukaemia-associated gingivitis
    - 2) Other
3. Gingival diseases modified by medications
  - a. Drug-influenced gingival diseases
    - 1) Drug-influenced gingival enlargements
    - 2) Drug-influenced gingivitis
      - a) Oral contraceptive-associated gingivitis
      - b) Other
4. Gingival diseases modified by malnutrition
  - a. Ascorbic acid-deficiency gingivitis
  - b. Other

TABLE 1

(continued)

**B. Non-plaque-induced gingival lesions**

1. Gingival diseases of specific bacterial origin
2. Gingival diseases of viral origin
  - a. Herpesvirus infections
    - 1) Primary herpetic gingivostomatitis
    - 2) Recurrent oral herpes
    - 3) Varicella-zoster infections
  - b. Other
3. Gingival diseases of fungal origin
  - a. Candida species infections
  - b. Linear gingival erythema
  - c. Histoplasmosis
  - d. Other
4. Gingival lesions of genetic origin
  - a. Hereditary gingival fibromatosis
  - b. Other
5. Gingival manifestations of systemic conditions
  - a. Mucocutaneous disorders
    - 1) Lichen planus
    - 2) Pemphigoid
    - 3) Pemphigus vulgaris
    - 4) Erythema multiforme
    - 5) Lupus erythematosus
    - 6) Drug-induced
    - 7) Other
  - b. Allergic reactions
    - 1) Dental restorative materials
    - 2) Reactions attributable to
      - a) Toothpastes/dentifrices
      - b) Mouthrinses/mouthwashes
      - c) Chewing gum additives
      - d) Foods and additives
    - 3) Other
6. Traumatic lesions (factitious, iatrogenic, accidental)
  - a. Chemical injury
  - b. Physical injury
  - c. Thermal injury
7. Foreign body reactions
8. Not otherwise specified (NOS)

**II. Chronic periodontitis****A. Localized****B. Generalized****III. Aggressive periodontitis****A. Localized****B. Generalized**



TABLE 1

(continued)

**IV. Periodontitis as a manifestation of systemic diseases****A. Associated with haematological disorders**

1. Acquired neutropenia
2. Leukaemias
3. Other

**B. Associated with genetic disorders**

1. Familial and cyclic neutropenia
2. Down syndrome
3. Leukocyte adhesion deficiency syndromes
4. Papillon-Lefèvre syndrome
5. Chediak-Higashi syndrome
6. Histiocytosis syndromes
7. Glycogen storage disease
8. Infantile genetic agranulocytosis
9. Cohen syndrome
10. Ehlers-Danlos syndrome (types IV and VIII)
11. Hypophosphatasia
12. Other

**C. Not otherwise specified (NOS)****V. Necrotizing periodontal diseases****A. Necrotizing ulcerative gingivitis (NUG)****B. Necrotizing ulcerative periodontitis (NUP)****VI. Abscesses of the periodontium****A. Gingival abscess****B. Periodontal abscess****C. Pericoronal abscess****VII. Periodontitis associated with endodontic lesions****A. Combined periodontic-endodontic lesions****VIII. Developmental or acquired deformities and conditions****A. Localized tooth-related factors that modify or predispose to plaque-induced gingival diseases/periodontitis**

1. Tooth anatomical factors
2. Dental restorations/appliances
3. Root fractures
4. Cervical root resorption and cemental tears

**B. Mucogingival deformities and conditions around teeth**

1. Gingival/soft tissue recession
2. Lack of keratinized gingiva
3. Decreased vestibular depth
4. Aberrant frenum/muscle position
5. Gingival excess
  - a. Pseudopocket
  - b. Inconsistent gingival margin
  - c. Excessive gingival display
  - d. Gingival enlargement (see I.A.3. and I.B.4.)
6. Abnormal colour

**C. Mucogingival deformities and conditions on edentulous ridges****D. Occlusal trauma**



## Reading List

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### Dental Plaque-Induced Gingivitis

- Definition** ▶ Dental plaque-induced gingivitis (chronic gingivitis) is gingivitis caused by the accumulation of dental bacterial plaque.
- Etiology** ▶ Accumulation of dental plaque on the tooth close to the gingivae.
- Gingival Involvement** ▶ Common; affects over 90% of the population.
- Other Sites of Involvements** ▶ None
- Main Clinical Features** ▶ Painless  
 ▶ May be bleeding from the gingival crevice, particularly when brushing  
 ▶ Gingival margins slightly red and swollen (**Figs. 13–16**)  
 ▶ Eventually mild gingival hyperplasia
- Diagnosis** ▶ Diagnosis is from history and clinical features.
- Differential Diagnosis** ▶ Desquamative gingivitis  
 ▶ Trauma  
 ▶ Drugs and plasma cell gingivostomatitis  
 ▶ Infections, e.g., primary herpetic stomatitis, candidiasis  
 ▶ Granulomas, e.g., Crohn's disease, orofacial granulomatosis or sarcoidosis  
 ▶ Erythroplasia  
 ▶ Kaposi's sarcoma
- Treatment** ▶ Plaque control  
 ▶ Where necessary, calculus removal by scaling and polishing

**FIGURE 13**

Plaque-induced gingivitis; early



FIGURE 14

Plaque-induced gingivitis; early



FIGURE 15

Plaque-induced gingivitis; advanced



FIGURE 16

Plaque-induced gingivitis; advanced





## Chronic Periodontitis

- Definition** ▶ Chronic periodontitis is inflammation of the gingivae and periodontal membrane (ligament).
- Etiology** ▶ Plaque accumulation and calcification to form calculus (tartar) above (supra-) and/or below (sub-gingival) the gum-line
- ▶ Anaerobes such as *Porphyromonas gingivalis*, *Bacteroides forsythus* and *Actinobacillus actinomycetemcomitans* are implicated
- ▶ The inflammatory reaction initiated by plaque is associated with progressive loss of periodontal ligament and alveolar bone and, eventually, tooth mobility and loss:
- The gingiva detaches from the tooth.
  - The periodontal membrane and alveolar bone are damaged.
  - An abnormal gap (pocket) develops between the tooth and gingiva.
  - Debris and pus may be expressed from the pockets (pyorrhoea).
- ▶ The teeth may slowly loosen and, without treatment, may eventually be lost.
- ▶ Subjects are rendered particularly susceptible by genetic and/or environmental factors including:
- Cigarette smoking
  - Interleukin-1 gene polymorphisms
  - Immune depression
  - Diabetes
  - Osteoporosis
- Periodontal Involvement** ▶ Exclusively
- Other Sites of Involvements** ▶ Studies are on-going to test possible effects of periodontal disease on systemic health and relationships with:
- Atherosclerosis
  - Hypertension
  - Coronary heart disease
  - Cerebrovascular disease
  - Low birth weight babies
  - Effects on diabetic control
- Main Clinical Features** ▶ Chronic Periodontitis
- Is typically seen in adults.
  - Is painless but may be associated with bleeding, halitosis (oral mal-odour) and a foul taste (**Figs. 17–21**).
  - May be a sequel of chronic gingivitis.
- Diagnosis** ▶ Diagnosis is from history and clinical features supplemented by:
- Periodontal probing
  - Imaging
- Differential Diagnosis** ▶ Other causes of tooth mobility including
- Trauma
  - Neoplasms
  - Immune defects
- Treatment** ▶ Improvement in oral hygiene
- ▶ Scaling and polishing
- ▶ Sometimes curettage
- ▶ Surgical removal of the pocket wall and removal of diseased tissue may be needed

FIGURE 17

Chronic periodontitis; early



FIGURE 18

Chronic periodontitis; early



FIGURE 19

Chronic periodontitis;  
moderately advanced

FIGURE 20

Chronic periodontitis;  
moderately advanced



FIGURE 21

Chronic periodontitis; advanced



### Aggressive Periodontitis

- Definition** ▶ This is where infection leads to more rapidly progressive periodontal disease.
- Etiology** ▶ Genetic variations in genes and gene products of cytokines and other inflammatory/destructive mediators appear to underlie early onset and severe types of periodontal disease, as well as specific genetic syndromes involving early periodontitis.
- ▶ The interleukin 1 (IL-1) genotype (IL-1A +4845 and IL-1B +3954) appears in about 30% of most Caucasian and African-American populations studied, and increases the risk of severe inflammatory periodontal disease by about 7 times.
- ▶ HLA serotypes may be associated with periodontal disease. The HLA-DRB1\*1501-DQB1\*0602 genotype has been found with increasing frequency in early onset periodontitis (EOP) and is associated with a strong T lymphocyte response against the *Porphyromonas gingivalis* antigen Ag53p141–161.



- ▶ Several other mechanisms apart from T cell responses may be involved in genetic susceptibility to periodontal disease, but particularly:
  - Inflammatory mediators such as interleukins, prostaglandins, tumor necrosis factor alpha (TNF- $\alpha$ ) and matrix metalloproteinases (MMPs)
  - Regulation of IgG subclass antibody responses
  - Polymorphonuclear neutrophilic leukocytes (PMNL) are essential for the phagocytosis and killing of a range of microorganisms, including periodontal pathogens
  - Periodontitis may develop despite good control of plaque and is then typically related to an inherited immune defect such as:
    - *Accelerated* (prepubertal) periodontitis is a rare condition seen in children.
    - *Localized juvenile* periodontitis is characterised by localized periodontal destruction, classically in the permanent incisor and first molar regions in adolescents or young adults in the absence of poor oral hygiene or gross systemic disease.
    - *Juvenile periodontitis* (periodontosis) is seen especially in females, and may be associated with minor defects of neutrophil function and with microorganisms such as *Actinobacillus actinomycetemcomitans* and capnocytophaga.
    - *Papillon-Lefèvre* syndrome, Down syndrome, type VIII Ehlers-Danlos syndrome, and hypophosphatasia.
  - An acquired immune defect such as in:
    - Poorly controlled diabetes mellitus
    - White cell dyscrasias including neutrophil defects and neutropenias
    - HIV/AIDS

**Periodontal Involvement** ▶ Exclusively

**Other Sites of Involvements** ▶ None unless there is recurrent infection in the immune defects

**Main Clinical Features** ▶ The features are those of marginal gingivitis and:

- Rapid destruction of alveolar bone support
- Deep pocket formation (**Figs. 22–24**)
- Tooth mobility
- Tooth migration
- Halitosis

**Diagnosis** ▶ Diagnosis is from history and clinical features  
 ▶ Diagnosis is supplemented by radiographical features

**Differential Diagnosis** ▶ A miscellany of immune disorders

**Treatment** ▶ Oral hygiene  
 ▶ Scaling and root planing  
 ▶ Periodontal surgery  
 ▶ Antimicrobials



FIGURE 22

Aggressive periodontitis



FIGURE 23

Aggressive periodontitis



FIGURE 24

Aggressive periodontitis



### Mouthbreathing Gingivitis

**Definition** ▶ Hyperplastic gingivitis related to mouthbreathing. The various factors that can cause a mouthbreathing habit include asthma, allergies and enlarged glandular tissue.

**Etiology** ▶ Mouthbreathing, especially nocturnally. Mouthbreathing, increased lip separation and decreased upper lip coverage at rest are all associated with higher levels of plaque and gingival inflammation.

**Gingival Involvement** ▶ Common

**Other Sites of Involvements** ▶ None

**Main Clinical Features** ▶ Enlarged gingivae, especially labially in the anterior maxilla (Figs. 25, 26)

FIGURE 25

Mouth breathing gingivitis



FIGURE 26

Mouth breathing gingivitis



**Diagnosis** ▶ Diagnosis is from history and clinical features

**Differential Diagnosis** ▶ Chronic gingivitis  
 ▶ Hereditary gingival fibromatosis  
 ▶ Drug-induced gingivitis  
 ▶ Pregnancy gingivitis  
 ▶ Sarcoidosis  
 ▶ Crohn's disease  
 ▶ Leukaemia  
 ▶ Wegener's granulomatosis  
 ▶ Amyloidosis  
 ▶ Scurvy  
 ▶ Mucopolysaccharidoses  
 ▶ Mucopolipidoses

**Treatment** ▶ Attention to nasal airway (ENT opinion)  
 ▶ Occasionally surgery

### Gingival Diseases of Specific Origin

#### Granulomatous Gingivitis

**Definition** ▶ Gingival swelling where lymphoedema and non-caseating granulomas may be seen.

**Etiology** ▶ It is unclear where in the spectrum of Crohn's disease/sarcoidosis/allergy/infection these lesions (and related conditions such as Melkersson-Rosenthal syndrome and granulomatous cheilitis) lie.  
 ▶ Some patients develop:

- Granulomatous reactions (orofacial granulomatosis) because of an adverse reaction to various food additives such as
  - Cinnamic aldehyde
  - Benzoates
  - Butylated hydroxyanisole
  - Dodecyl gallate (in margarine)
  - Menthol (in peppermint oil)
- Crohn's disease after some time

**Gingival Involvement** ▶ Rare; purple granulomatous enlargements may appear on the gingiva.

**Other Sites of Involvements** ▶ Lips or face

**Main Clinical Features** ▶ Onset is usually in young adult life  
 ▶ Earliest manifestation is sudden diffuse or occasionally nodular non-tender swellings of the lip (cheilitis granulomatosa) or face  
 ▶ Granulomatous gingiva enlargements (Figs. 27, 28)  
 ▶ Lingual, palatal, and buccal involvement also may occur  
 ▶ Mucosal lesions include thickening and folding to produce:
 

- A "cobblestone" type of appearance
- Mucosal tags.

 ▶ Linear ulcers in the vestibule, often with granulomatous masses flanking them, classically involve the buccal sulcus



FIGURE 27

Granulomatous gingivitis



FIGURE 28

Granulomatous gingivitis



- Diagnosis**
- ▶ Diagnosis is from history and clinical features supplemented by
  - ▶ Histopathology
  - ▶ Investigation to exclude Crohn's disease (endoscopy, imaging and histopathology of the gastrointestinal tract)
  - ▶ Investigations to exclude sarcoidosis (chest imaging, serum angiotensin converting enzyme, and gallium scan)
  - ▶ Patch tests to exclude reactions to foodstuffs or additives.

- Differential Diagnosis**
- ▶ Chronic gingivitis
  - ▶ Desquamative gingivitis
  - ▶ Trauma
  - ▶ Drugs and plasma cell gingivostomatitis
  - ▶ Infections e.g. primary herpetic stomatitis, candidiasis
  - ▶ Erythroplasia
  - ▶ Kaposi's sarcoma
  - ▶ Hyperplastic gingivitis
  - ▶ Hereditary gingival fibromatosis



- ▶ Pregnancy
- ▶ Leukaemia
- ▶ Wegener's granulomatosis
- ▶ Amyloidosis
- ▶ Scurvy
- ▶ Mucopolysaccharidoses
- ▶ Mucopolipidoses

- Treatment**
- ▶ Possible provoking dietary components should avoided
  - ▶ Drug management has included:
    - Clofazimine
    - Nonsteroidal antiinflammatory agents
    - Antibiotics
    - Antimalarials
    - Mast cell stabilizers
    - Intralesional corticosteroid injections

### "Geographic" Gingivitis

- Definition**
- ▶ Erythema migrans (geographic stomatitis, benign migratory glossitis, stomatitis areata migrans)
    - Typically affects the tongue
    - Is a benign inflammatory condition unrelated to cutaneous erythema migrans.
    - Is common, affecting about 1–2% of the population

- Etiology**
- ▶ The aetiology is unknown
  - ▶ There is a genetic background
    - A positive family history may be obtainable.
    - HLA findings have been equivocal, with reports of associations with B15 and DR7.
    - Many patients have a fissured tongue (plicated tongue)
  - ▶ Some patients
    - Have atopic allergies such as hay fever
    - Relate the lesions to a particular food e.g. cheese
    - Relate the lesions to stress.
    - Have psoriasis, mainly the severe pustular variety

- Gingival Involvement**
- ▶ Rare

- Other Sites of Involvements**
- ▶ The tongue is usually, but not invariably, affected

- Main Clinical Features**
- ▶ Rounded, sometimes scalloped, map-like reddish areas with a white margin
  - ▶ The patterns change from day to day and even over a few hours (Figs. 29–31)
  - ▶ It may be asymptomatic or sore

- Diagnosis**
- ▶ Diagnosis is from history and clinical features supplemented by
  - ▶ Blood and urine examination to exclude anaemia and diabetes.

- Differential Diagnosis**
- ▶ Lichen planus
  - ▶ Candidosis
  - ▶ Deficiency glossitis
  - ▶ Generalized pustular psoriasis

FIGURE 29

Geographic gingivitis



FIGURE 30

Geographic tongue



FIGURE 31

Geographic stomatitis



- ▶ Reiter's syndrome
- ▶ Syphilis
- ▶ Ulcerative colitis
- ▶ Acrodermatitis continua of Hallopeau
- ▶ Larva migrans

- Treatment**
- ▶ Reassurance.
  - ▶ Topical corticosteroids may be employed for severely symptomatic patients.

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### Torus Mandibularis

- Definition** ▶ A bilateral asymptomatic benign bony lump or lumps, lingual to the mandibular premolars
- Etiology** ▶ Developmental  
 ▶ Seen especially in Asian and Black races  
 ▶ May be associated with parafunction  
 ▶ Seen mainly in males, and rarely in Turner's syndrome (4S,X)
- Gingival Involvement** ▶ Common
- Other Sites of Involvement** ▶ Often associated with torus palatinus; a slow-growing, asymptomatic, benign bony lump in midline of palate
- Main Clinical Features** ▶ Bony lumps, which grow slowly, appear in adolescence and usually cease growth in late teens (**Fig. 32**).  
 ▶ Bony hard with normal overlying mucosa, asymptomatic, benign and of no consequence.
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:  
 • Radiographic examination.
- Differential Diagnosis** ▶ Torus mandibularis – exclude unerupted teeth  
 ▶ Torus palatinus – exclude neoplasms
- Treatment** ▶ Excise or reduce only if causing severe difficulties with dentures.

**FIGURE 32**

Torus mandibularis



### Multiple Exostoses of the Maxillary Alveolar Process

- Definition** ▶ Multiple exostoses of the maxillary alveolar process
- Etiology** ▶ Genetic
- Gingival Involvement** ▶ Rare
- Other Sites of Involvement** ▶ Occasionally arise beneath a bridge pontic (sub-pontic osseous hyperplasia)
- Main Clinical Features** ▶ Exostoses typically buccal to maxillary premolars and molars (Figs. 33, 34).  
 ▶ Appear in adolescence.  
 ▶ Grow slowly and usually cease growth in late teens.  
 ▶ Most are asymptomatic, benign and of no consequence.
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:  
 • Radiography.
- Differential Diagnosis** ▶ Osteoma
- Treatment** ▶ Leave alone

### Fibrous Tuberosity

- Definition** ▶ A fibrous tuberosity
- Etiology** ▶ Idiopathic
- Gingival Involvement** ▶ Uncommon
- Other Sites of Involvement** ▶ None
- Main Clinical Features** ▶ Enlarged tuberosities, mainly palatally (Figs. 35–37)
- Diagnosis** ▶ Diagnosis is from history and clinical features.
- Differential Diagnosis** ▶ Hereditary gingival fibromatosis  
 ▶ Drugs  
 ▶ Neoplasms  
 ▶ Pregnancy gingivitis  
 ▶ Sarcoidosis  
 ▶ Crohn's disease  
 ▶ Leukaemia
- Treatment** ▶ Occasionally surgery

**FIGURE 33**

Multiple exostoses

**FIGURE 34**

Multiple exostoses

**FIGURE 35**

Fibrous tuberosity



FIGURE 36

Fibrous tuberosity



FIGURE 37

Fibrous tuberosity



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## Drug-Influenced Gingival Lesions and Diseases

### Gingival Enlargement Due to Phenytoin

**Definition** ► Gingival swelling in response to the use of phenytoin

**Etiology** ► Exact mechanism unknown  
 ► Positive correlation between the severity of gingival overgrowth and:

- Inflammation
- Plaque score
- Calculus accumulation
- Pocket depths

**Gingival Involvement** ► Common

**Other Sites of Involvement** ► This may be associated with hypertrichosis.

**Main Clinical Features** ► Gingival enlargement (**Figs. 38, 39**). Characteristically is:

- Firm, painless, pale, tough with coarse stippling and relatively little tendency to bleed. It is seen:
  - On the papillae mainly.
  - On buccal and labial gingiva more than palatal and lingual gingiva.
  - Not in edentulous sites.
  - First 2–3 months after treatment is started.
  - Peaking at around 1 year of drug use, it is aggravated by poor oral hygiene.

**FIGURE 38**

Drug-induced gingival swelling due to phenytoin



FIGURE 39

Drug-induced gingival swelling  
due to phenytoin



**Diagnosis** ▶ Diagnosis is from history and clinical features.

**Differential Diagnosis** ▶ Chronic gingivitis  
▶ Hyperplastic gingivitis  
▶ Hereditary gingival fibromatosis  
▶ Drugs  
▶ Pregnancy gingivitis  
▶ Sarcoidosis  
▶ Crohn's disease  
▶ Leukaemia  
▶ Wegener's granulomatosis  
▶ Amyloidosis  
▶ Scurvy  
▶ Mucopolysaccharidoses  
▶ Mucopolipidoses

**Treatment** ▶ In consultation with physician, change or reduce dose of the drug.  
▶ Improve oral hygiene.  
▶ Gingival surgery may be needed.

### Gingival Enlargement Due to Calcium Channel Blockers

**Definition** ▶ Gingival swelling in response to use of calcium channel blockers

**Etiology** ▶ Calcium channel blockers, mainly the dihydropyridines such as nifedipine used as antihypertensives, can produce gingival hyperplasia similar to that induced by phenytoin.  
▶ Exact mechanism is unknown.

**Gingival Involvement** ▶ Typically affects the papillae, which become red and puffy and tend to bleed.

**Other Sites of Involvement** ▶ Hypertrichosis

FIGURE 40

Drug-induced gingival swelling  
due to nifedipine



FIGURE 41

Drug-induced gingival swelling  
due to nifedipine



FIGURE 42

Drug-induced gingival swelling  
due to nifedipine





- Main Clinical Features**
- ▶ Changes often develop slowly – over weeks rather than days, usually painless and aggravated by poor oral hygiene.
  - ▶ Starts interdentally, especially labially.
  - ▶ Papillae are firm, pale and enlarge to form false vertical clefts (Figs. 40–42).

- Diagnosis**
- ▶ Diagnosis is from history and clinical features.

- Differential Diagnosis**
- ▶ Chronic gingivitis
  - ▶ Hyperplastic gingivitis
  - ▶ Hereditary gingival fibromatosis
  - ▶ Drugs
  - ▶ Pregnancy gingivitis
  - ▶ Sarcoidosis
  - ▶ Crohn's disease
  - ▶ Leukaemia
  - ▶ Wegener's granulomatosis
  - ▶ Amyloidosis
  - ▶ Scurvy
  - ▶ Mucopolysaccharidoses
  - ▶ Mucopolipidoses

- Treatment**
- ▶ In consultation with a physician, change or reduce dose of the drug.
  - ▶ Improve oral hygiene.
  - ▶ Gingival surgery may be needed.

### Gingival Enlargement Due to Ciclosporin

- Definition**
- ▶ Gingival swelling in response to use of ciclosporin

- Etiology**
- ▶ Exact mechanism unknown
  - ▶ Usually aggravated by poor oral hygiene

- Gingival Involvement**
- ▶ *Ciclosporin* (cyclosporine), an immunosuppressive drug, can cause gingival swelling, but only a third of patients may be affected, more commonly children.

- Other Sites of Involvement**
- ▶ Hypertrichosis

- Main Clinical Features**
- ▶ Ciclosporin hyperplasia closely resembles that induced by phenytoin.
  - ▶ It is seen mainly anteriorly and labially.
  - ▶ It is exacerbated by poor oral hygiene and concurrent administration of nifedipine.
  - ▶ It appears not to be proportional to the drug dose used.
  - ▶ Papillae are firm, pale and enlarge to form false vertical clefts (Figs. 43–47).
  - ▶ Changes develop slowly – over weeks rather than days – and are usually painless.

- Diagnosis**
- ▶ Diagnosis is from history and clinical features.

- Differential Diagnosis**
- ▶ Chronic gingivitis
  - ▶ Hyperplastic gingivitis
  - ▶ Hereditary gingival fibromatosis
  - ▶ Drugs



**FIGURE 43**

Drug-induced gingival swelling  
due to ciclosporin

**FIGURE 44**

Drug-induced gingival swelling  
due to ciclosporin

**FIGURE 45**

Drug-induced gingival swelling  
due to ciclosporin



FIGURE 46

Drug-induced gingival swelling  
due to ciclosporin



FIGURE 47

Drug-induced gingival swelling  
due to ciclosporin



- ▶ Pregnancy gingivitis
- ▶ Sarcoidosis
- ▶ Crohn's disease
- ▶ Leukaemia
- ▶ Wegener's granulomatosis
- ▶ Amyloidosis
- ▶ Scurvy
- ▶ Mucopolysaccharidoses
- ▶ Mucopolipidoses

**Treatment**

- ▶ In consultation with a physician, change or reduce dose of the drug.
- ▶ Improve oral hygiene.
- ▶ Gingival surgery may be needed.

### Plasma Cell Gingivitis (Idiopathic Circumoral Plasmacytosis)

**Definition** ▶ Plasma-cell gingivitis (PCG) is a rare, non-symptomatic erythematous and oedematous reaction to various substances, found in the attached gingiva.

**Etiology** ▶ Chewing gum additives such as:

- Cinnamonaldehyde
- Cinnamon oil
- Dichlorophene
- Hexylresorcinol
- Mint

**Gingival Involvement** ▶ Uncommon

**Other Sites of Involvement** ▶ Rarely, other oral or pharyngeal sites. Lesions with similar clinical appearance are also found in the genital mucosa of patients with a PCG.

FIGURE 48

Plasma-cell gingivitis



FIGURE 49

Plasma-cell gingivitis





FIGURE 50

Plasma-cell gingivitis



FIGURE 51

Plasma-cell gingivitis



- Main Clinical Features** ▶ Clinically, PCG is a sharply demarcated red area within the attached gingiva (Figs. 48–51).
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:
- Histopathology; abundant plasma cells are found in the connective tissue beneath a thin epithelium.
- Differential Diagnosis** ▶
- ▶ Chronic gingivitis
  - ▶ Hyperplastic gingivitis
  - ▶ Hereditary gingival fibromatosis
  - ▶ Drugs
  - ▶ Pregnancy gingivitis
  - ▶ Sarcoidosis
  - ▶ Crohn's disease
  - ▶ Leukaemia
  - ▶ Wegener's granulomatosis
  - ▶ Amyloidosis



- ▶ Scurvy
- ▶ Mucopolysaccharidoses
- ▶ Mucolipidoses

**Treatment** ▶ Corticosteroids

### Allergic Gingivitis

**Definition** ▶ An uncommon erythematous and oedematous reaction to various substances

- Etiology** ▶ Constituents of perfumes, sprays, creams, mouthwashes, dentifrices and breath fresheners:
- Furocoumarin
  - Bergamot oil
  - Eugenol
  - Others
- ▶ A wide range of other allergens:
- Food and additives
  - Natural products
  - Chewing gum additives
  - Dental materials
  - Rubber products, such as dental dam or impression materials
  - Silicone-based impression materials
  - Eugenol and other essential oils
  - Mercury
  - Nickel
  - Beryllium
  - Acrylic resin components
  - Epoxy resin components
  - Drugs

**Gingival Involvement** ▶ Uncommon

**Other Sites of Involvement** ▶ Various

**Main Clinical Features** ▶ Swollen red gingivae (**Figs. 52–54**)

- Diagnosis** ▶ Diagnosis is from history and clinical features. A careful history is required, concentrating on the use of chewing gum or oral medications.
- ▶ Patch tests on the skin are usually negative.

- Differential Diagnosis** ▶ Chronic gingivitis
- ▶ Desquamative gingivitis
- ▶ Trauma
- ▶ Drugs
- ▶ Infections, e.g. primary herpetic stomatitis, candidiasis
- ▶ Granulomas, e.g. Crohn's disease, orofacial granulomatosis or sarcoidosis
- ▶ Erythroplasia
- ▶ Kaposi's sarcoma

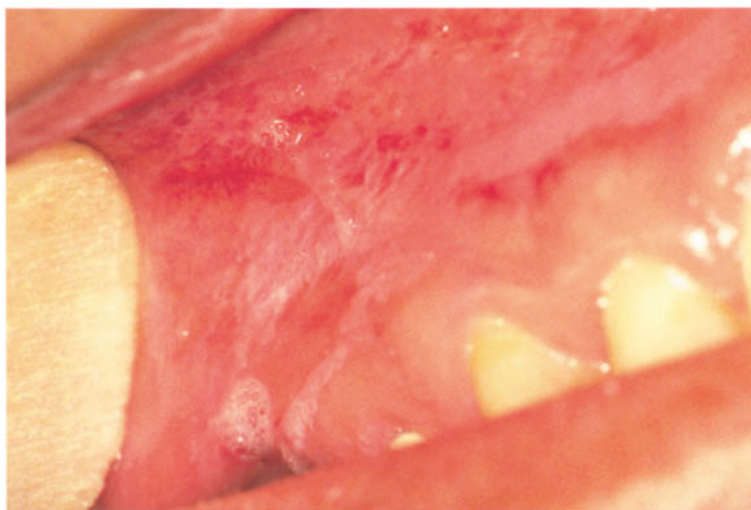
- Treatment** ▶ Withdraw the offending agent.
- ▶ Corticosteroids occasionally.

**FIGURE 52**

Drug-induced lesion  
due to hydroxyurea

**FIGURE 53**

Drug-induced lesion  
due to sanguinaria

**FIGURE 54**

Drug-induced lesion  
due to sanguinaria



### Heavy Metal Deposition and Other Causes of Hyperpigmentation

**Definition** ▶ Hyperpigmentation induced by habits, drugs or other chemicals

- Etiology**
- ▶ Betel (Areca) nut.
  - ▶ Smoking.
  - ▶ Drugs:
    - Phenytoin.
    - Antimalarials.
    - ACTH.
    - Clofazimine.
    - Contraceptive pill.
    - Zidovudine.
    - Ketoconazole.
    - Busulphan.
    - Minocycline.
    - Phenothiazines.
  - ▶ Heavy metals, rarely:
    - Amalgam.
    - Arsenic.
    - Bismuth.
    - Copper.
    - Gold.
    - Lead.
    - Mercury.
    - Platinum.
    - Silver.
    - Zinc.
  - ▶ Staining of gingivae adjacent to crowns or other restorative dental materials appears due mainly to silver, sulphur and selenium.
  - ▶ Rare cases of toxicity are related to use of platinum in cancer therapy.
  - ▶ Many heavy metals formerly implicated are not used now therapeutically.
  - ▶ Cases of heavy metal toxicity related to industrial exposure are still occasionally seen.
  - ▶ Metal sulphides form due to the activity of plaque microorganisms and produce pigmentation.

**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ Any oral location  
▶ Occasionally skin, nails or other sites

- Main Clinical Features**
- ▶ Hyperpigmentation (sometimes called Burton's line): brown or blue or black line, mainly at gingival margin and particularly in inflamed sites (Figs. 55–57).
  - ▶ Sometimes heavy metal poisoning may result in tooth loss.
  - ▶ Rarely, cobalt may induce orofacial granulomatosis.
  - ▶ Patchy mucosal hyperpigmentation with, in smokers or betel users, tooth staining.

- Diagnosis**
- ▶ Diagnosis is from history and clinical features, supplemented by:
    - Histopathology in some instances.
  - ▶ Imaging may be helpful whenever hyperpigmentation may be caused by amalgam, graphite or a foreign body.
  - ▶ Photographs may be useful for future comparison.



**FIGURE 55**

Drug-induced gingival lesions  
due to bismuth

**FIGURE 56**

Drug-induced gingival lesions  
due to lead

**FIGURE 57**

Betel nut staining





- Differential Diagnosis**
- ▶ Racial pigmentation
  - ▶ Localized irritation, e.g. smoking
  - ▶ Amalgam tattoo
  - ▶ Naevus
  - ▶ Melanotic macule
  - ▶ Malignant melanoma
  - ▶ Kaposi's sarcoma
  - ▶ Drugs, e.g. antimalarials
  - ▶ Addison's disease
- Treatment**
- ▶ Remove the cause.
  - ▶ Treatment of any heavy metal poisoning.
  - ▶ Reassurance.

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**Gingival Abscess**

- Definition** ▶ An abscess in the gingiva
- Etiology** ▶ Usually a foreign body such as dental floss or particle of food or dental restorative material
- Gingival Involvement** ▶ Rare
- Main Clinical Features** ▶ Pain, swelling and erythema (**Fig. 58**)
- Diagnosis** ▶ Diagnosis is from history and clinical features.
- Differential Diagnosis** ▶ Periodontal abscess  
▶ Periapical abscess
- Treatment** ▶ Drainage

**FIGURE 58**

Gingival abscess



FIGURE 59

Periodontal abscess



FIGURE 60

Periodontal abscess



FIGURE 61

Periodontal abscess





### Periodontal Abscess

- Definition** ▶ An abscess arising in a periodontal pocket
- Etiology** ▶ Seen almost exclusively in patients with chronic periodontitis.  
▶ Debris and pus cannot escape easily from the periodontal pocket and therefore an abscess results.  
▶ May follow impaction of a foreign body.  
▶ Rarely related to a lateral root canal on a non-vital tooth.
- Gingival Involvement** ▶ Uncommon
- Other Sites of Involvement** ▶ None
- Main Clinical Features** ▶ Lateral periodontal abscesses usually present with:  
• Pain  
• Swelling  
▶ Discharge either through the pocket or buccally, but more coronally than a periapical abscess (**Figs. 59–61**)
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:  
• Imaging.
- Differential Diagnosis** ▶ Gingival abscess  
▶ Periapical abscess  
▶ Neoplasm
- Treatment** ▶ Establish drainage, usually by curettage

### Pericoronal Abscess

- Definition** ▶ An abscess in the pericoronal region of an impacted tooth, typically a mandibular third molar
- Etiology** ▶ Trauma  
▶ Plaque accumulation  
▶ Necrotizing gingivitis  
▶ A foreign body such as food  
▶ Impaired immunity such as in leukaemia or infectious mononucleosis
- Gingival Involvement** ▶ Rare
- Other Sites of Involvement** ▶ None
- Main Clinical Features** ▶ Pain, swelling (**Fig. 62**), erythema and trismus
- Diagnosis** ▶ Diagnosis is from history and clinical features.
- Differential Diagnosis** ▶ Periodontal abscess  
▶ Periapical abscess
- Treatment** ▶ Drainage  
▶ Antimicrobials  
▶ Surgical opinion on the partially erupted tooth

FIGURE 62

Pericoronal abscess



### Tooth Abscess

**Definition** ▶ An abscess around the apex of a non-vital tooth

**Etiology** ▶ Untreated, dental caries can progress through dentine to the pulp which becomes inflamed (pulpitis) producing severe persistent pain (toothache). The pulp undergoes necrosis when inflammation can spread around the tooth apex (periapical periodontitis), forming an abscess (periapical, or dental, abscess).  
 ▶ Occasionally a dental abscess arises from:

- A necrotic pulp in a tooth damaged by trauma.
- A foreign body.
- Unusual oral infections such as actinomycosis, nocardiosis or botryomycosis.

**Gingival Involvement** ▶ Common

**Other Sites of Involvement** ▶ None

**Main Clinical Features** ▶ Most abscesses present with:

- Pain, and swelling (Figs. 63, 64).
- Sometimes fever and/or cervical lymph node enlargement.

▶ Most abscesses discharge in the mouth on the buccal gingiva but occasionally they discharge palatally or lingually or on the chin or submental region or elsewhere (Figs. 65–67).

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Vitality testing.
- Radiographic imaging.

**Differential Diagnosis** ▶ Periodontal abscess  
 ▶ Rarely, metastasis

**Treatment** ▶ Dental abscesses are drained by tooth extraction, incision and drainage, or through the root canal (endodontics). Antimicrobials may be indicated.

**FIGURE 63**

Periapical abscess

**FIGURE 64**

Periapical abscess

**FIGURE 65**

Fistula





FIGURE 66

Fistula



FIGURE 67

Fistula with pigmentation



### Oroantral Fistula

- Definition** ▶ A fistula is a non-anatomical epithelial lined tract joining two epithelial surfaces. An oro-antral fistula (OAF) is between the mouth (lined with squamous epithelium) and the maxillary antrum (lined with a ciliated columnar epithelium with mucous glands: respiratory epithelium).
- Etiology** ▶ As the sinus enlarges with growth it approaches closer to the teeth and the risk of creating an OAF when removing upper teeth, or of displacing teeth or roots into the antrum, is therefore much greater in adults. OAF is most often caused by the extraction of maxillary teeth, especially the molars or premolars, closely related to the antrum. Major fistulae tend to occur if the maxillary tuberosity is fractured, particularly if several teeth are attached.
- ▶ Persistent OAFs tend to occur when a molar tooth root is accidentally displaced into the maxillary sinus or when a molar tooth is extracted.



FIGURE 68

Oroantral fistula



One situation that is especially prone to fistula formation is the extraction of a single standing maxillary molar or premolar whose roots are surrounded both posteriorly and anteriorly by the maxillary antrum. Roots of any maxillary premolar or molar tooth may be displaced into the antrum but the complete tooth most commonly displaced into the antrum is the unerupted upper third molar. On the other hand, small OAFs occur most frequently with extraction of the first or second molar.

- ▶ Rarely, facial trauma, neoplasms or osteomyelitis of the maxilla and surgery to the maxilla may lead to fistula formation.

**Gingival Involvement** ▶ Rare

- Main Clinical Features**
- ▶ Signs and symptoms from an OAF may occur immediately after the extraction of a tooth or may take some weeks to develop (Fig. 68).
  - ▶ In the early post-extraction period, there may be ipsilateral nose bleed (epistaxis).
  - ▶ The most common symptom is reflux of fluid from the mouth into the nose when the patient drinks. Fluid then dribbles from the nose.
  - ▶ A little later, as sinusitis develops, an unpleasant ipsilateral nasal discharge may arise.

- Diagnosis**
- ▶ Diagnosis is from history and clinical features. An obvious OAF may be seen in the mouth but often a small hole cannot be seen.
  - ▶ A way of testing for an established fistula is to place a small cotton wool pledget in the area, and ask the patient to squeeze their nose and then to attempt to blow it. This will force the cotton wool out of the fistula, and bubbles of air into the mouth through the fistula. Failing this, very gentle probing with a blunt probe, e.g. a lacrimal duct dilator, will reveal the fistula but if it is suspected that a fistula has been newly created, it is better to assume the presence of an OAF and conduct a repair without such testing.
  - ▶ Sometimes a red lump is noticed in the region of an OAF. This is prolapsed antral mucosa or occasionally a prolapsed antral polyp or mucosal cyst. Sometimes a fairly large fistula may be temporarily closed by

a prolapse of the antral mucosa. A blunt probe will push back the prolapsed tissue and reveal an obvious fistula.

- ▶ Symptoms of sinusitis may develop later but, in approximately one-quarter of patients with a fistula, there are no symptoms. This is usually because the fistula is so small that it is closed off by surrounding mucosa, particularly if it is in the buccal sulcus where the cheek may lie against it.

#### Differential Diagnosis ▶ Oronasal fistula

- Treatment**
- ▶ The treatment indicated depends on the rapidity with which the diagnosis of OAF has been established.
  - ▶ If diagnosis has been made as soon as fistula has been created – assuming that a tooth or tooth root has not been displaced into the antrum – the immediate treatment is to close the tooth socket. For the experienced practitioner, the procedure is to raise a buccal flap and advance it over the fistula. The less experienced practitioner should simply suture the socket. The patient should then be given prophylactic treatment against maxillary sinusitis which would otherwise delay healing of the fistula; that is:
    - The patient should avoid blowing their nose.
    - Antibiotics such as erythromycin or co-trimoxazole, nose drops (ephedrine 0.5% or xylometazoline hydrochloride 0.1%) and inhalations (such as menthol and benzoin inhalation BP or menthol and eucalyptus inhalation BP qds) should be used.
  - ▶ Up to 48 h after the tooth extraction, an OAF can be repaired by advancement of a buccal flap but prophylactic antibiotics are indicated.

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**Self-Inflicted Injury**

- ▶ Abrasive toothbrushing
- ▶ Chewing on sticks
- ▶ Self-induced wounds
- ▶ Tattooing

- Etiology**
- ▶ Most gingival physical damage is from abrasive toothbrushing, usually over-vigorous horizontal brushing.
  - ▶ Children may damage their gingiva with a fingernail.
  - ▶ Gingival recession may be induced by oral jewelry such as tongue studs.
  - ▶ Gingival damage may be seen in:
    - Learning disability.
    - Lesch-Nyhan syndrome.
    - Syndromes with pain insensitivity:
      - Congenital indifference to pain.
      - Riley Day syndrome.

- Gingival Involvement** ▶ Uncommon

- Other Sites of Involvement** ▶ Any

- Main Clinical Features** ▶ Damage to the gingival margin or other areas accessible to the dominant hand usually (**Figs. 69–73**).

- Diagnosis** ▶ Diagnosis is from history and clinical features.

- Differential Diagnosis**
- ▶ Neoplasms
  - ▶ Aphthae
  - ▶ Systemic disease
    - Infections
    - Haematological disease
    - Gastrointestinal disease
    - Mucocutaneous disease
    - Drugs

- Treatment**
- ▶ Psychotherapy
  - ▶ Protective splints



FIGURE 69

Gingival trauma: toothbrushing



FIGURE 70

Gingival trauma: toothbrushing



FIGURE 71

Gingival trauma:  
hyperplasia due to toothbrushing





FIGURE 72

Gingival trauma: from a fingernail



FIGURE 73

Gingival trauma: from a fingernail



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**Definition** ▶ Chemically induced lesions often present as mucosal burns or white lesions.

**Etiology** ▶

- ▶ **Drugs**
  - Analgesic tablets
  - Cocaine deliberately rubbed into the gingivae or vestibule
  - Pancreatin can cause ulceration
- ▶ **Mouthwashes**
  - Chlorhexidine
  - Others, especially alcohol-containing washes
- ▶ **Dental procedures**
  - Acids (chromic, trichloracetic, phosphoric)
  - Self-curing resins, especially epoxy resins
- ▶ **Natural products**
  - Tobacco products including smokeless tobacco
  - Sanguinaria
  - Areca nut
  - The houseplant Dieffenbachia
  - The enzyme bromelin in pineapple
  - Others

**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ Frequent

**Main Clinical Features** ▶ Mucosal burns or white lesions (**Figs. 74–80**)

**Diagnosis** ▶ Diagnosis is from history and clinical features.

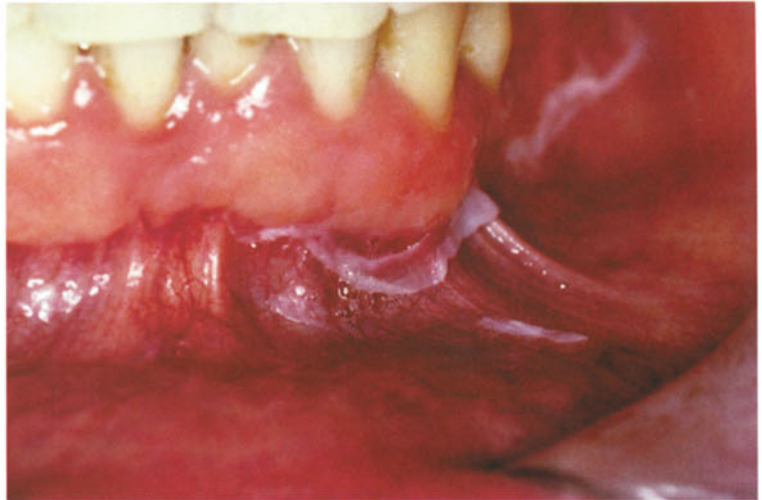
**Differential Diagnosis** ▶

- ▶ Materia alba
- ▶ Keratoses
- ▶ Carcinoma
- ▶ Candidiasis (thrush: candidal leukoplakia)
- ▶ Epstein-Barr virus (hairy leukoplakia)
- ▶ Syphilis (syphilitic mucous patches and keratosis)
- ▶ Lichen planus
- ▶ Lupus erythematosus

**Treatment** ▶ Remove the offending agent

**FIGURE 74**

Mucosal burn:  
chemical – from a mouthwash

**FIGURE 75**

Mucosal burn:  
chemical – from aspirin

**FIGURE 76**

Mucosal burn:  
chemical – from hydrogen peroxide





**FIGURE 77**

Mucosal burn:  
chemical – from trichloroacetic acid

**FIGURE 78**

Mucosal burn:  
chemical – from alcohol

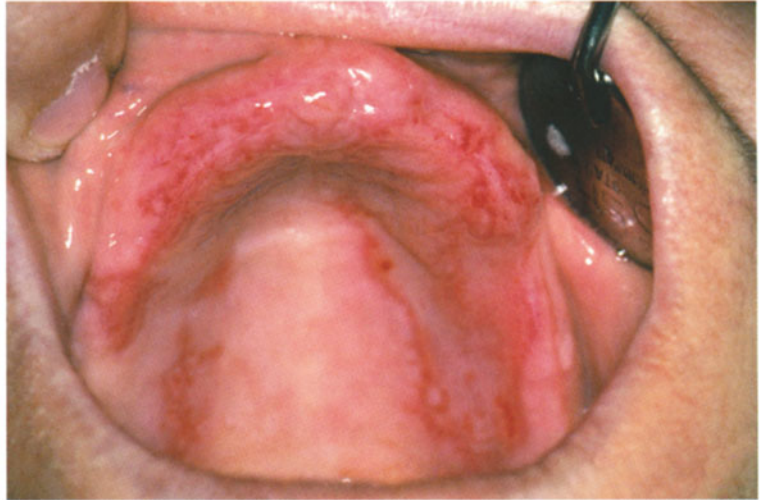
**FIGURE 79**

Mucosal burn:  
chemical – from iodine



FIGURE 80

Mucosal burn:  
chemical – from acrylic monomer



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- Definition** ▶ Thermally induced lesions often present as mucosal burns or white lesions.
- Etiology** ▶ Hot foods  
▶ Hot instruments (dental handpiece or extraction forceps usually)  
▶ Electrical burns  
▶ Cryosurgery
- Gingival Involvement** ▶ Rare
- Other Sites of Involvement** ▶ Seen especially on the palate or tongue, for example, so-called pizza-palate
- Main Clinical Features** ▶ White lesions, blisters or ulcers (**Fig. 81**)
- Diagnosis** ▶ Diagnosis is from history and clinical features.
- Differential Diagnosis** ▶ Materia alba  
▶ Burns  
▶ Keratoses  
▶ Carcinoma  
▶ Candidiasis (thrush: candidal leukoplakia)  
▶ Epstein-Barr virus (hairy leukoplakia)  
▶ Syphilis (syphilitic mucous patches and keratosis)  
▶ Lichen planus  
▶ Lupus erythematosus
- Treatment** ▶ Remove the offending agent.  
▶ Supportive care.

**Mucositis**

- Definition** ▶ Radiotherapy involving the mouth invariably produces burns and mucositis and if it involves the salivary glands, often produces xerostomia.
- Etiology** ▶ Ionizing radiation
- Gingival Involvement** ▶ Uncommon
- Other Sites of Involvement** ▶ Irradiation can also produce:
- Xerostomia, which predisposes to:
    - Dental caries.
    - Candidiasis.
  - Osteoradionecrosis.
  - Scarring is later followed by the appearance of telangiectasia.
  - Taste loss.
  - Subsequent neoplasia: for example, radiotherapy to oropharyngeal neoplasms predisposes to subsequent salivary neoplasms.
- Main Clinical Features** ▶ Diffuse erythema and ulceration (**Figs. 82–83**).  
 ▶ Gingival parameters such as bleeding index, plaque index and gingival recession are all increased after radiotherapy.
- Diagnosis** ▶ Diagnosis is from history and clinical features.
- Differential Diagnosis** ▶ Burns  
 ▶ Lichen planus  
 ▶ Inflammatory gingivitis  
 ▶ Desquamative gingivitis  
 ▶ Herpetic stomatitis, candidiasis  
 ▶ Granulomas, e.g. Crohn's disease, orofacial granulomatosis or sarcoidosis
- Treatment** ▶ There is no specific treatment.  
 ▶ Sucking ice is as satisfactory as any of the multitude of drugs and other agents available.  
 ▶ Analgesics



FIGURE 82

Mucosal burn:  
radiation



FIGURE 83

Mucosal burn:  
radiation mucositis



### Osteoradionecrosis

- Definition** ▶ Bone necrosis and infection arising in a previously irradiated jaw
- Etiology** ▶ Radiotherapy involving the jaws, especially the mandible, may predispose to osteoradionecrosis.  
▶ Endarteritis obliterans decreases the vascular supply to bone, predisposing to osteoradionecrosis.  
▶ The initiating factor is often trauma, such as tooth extraction or infection.  
▶ Osteoradionecrosis used to be a common complication of radiotherapy for head and neck cancers but modern techniques have significantly reduced the risk.
- Gingival Involvement** ▶ Rare

FIGURE 84

Osteoradionecrosis



- Other Sites of Involvement**
- ▶ Muscle involvement can produce trismus.
  - ▶ Developing teeth can be involved to cause:
    - Hypoplasia.
    - Stunted root formation.
    - Retarded eruption.
  - ▶ The mandibular condyle, or other growth areas, can be damaged resulting in facial deformity.
- Main Clinical Features**
- ▶ Presentation is of exposed bone in an irradiated mouth with or without:
    - External sinuses (**Fig. 84**).
    - Pain.
    - Pathological fracture.
  - ▶ Once established, the process may spread to involve the whole jaw, usually the mandible.
- Diagnosis**
- ▶ Diagnosis is from history and clinical features.
  - ▶ Microbiology.
  - ▶ Imaging.
  - ▶ Others.
- Differential Diagnosis**
- ▶ Osteomyelitis
- Treatment**
- ▶ Long-term antibiotics, especially tetracycline.
  - ▶ Local cleansing with aqueous 0.2% chlorhexidine.
  - ▶ Sequestrectomy and surgical intervention should be minimal.
  - ▶ Resection only as a last resort.

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## Pigmented Disorders

### Physiological Pigmentation

- Definition** ▶ Pigmentation related to race (racial pigmentation)
- Etiology** ▶ The most usual cause of brown oral mucosal pigmentation is ethnic, mainly in descendants of Blacks, but may be seen even in some fairly light-skinned people.
- Gingival Involvement** ▶ Common, especially anteriorly and labially
- Other Sites of Involvement** ▶ Many other sites may be affected but pigmentation is most prominent on the gingivae, buccal mucosa and tongue.
- Main Clinical Features** ▶ Brown macular areas, most obvious in anterior labial gingivae (**Figs. 85, 86**), mainly on attached gingivae
- Diagnosis** ▶ For generalized or multiple hyperpigmentation which is not clearly racial:
- Blood pressure should be taken to exclude the hypotension of Addison's disease.
  - Blood tests may be needed if there is suspicion of Addison's disease; plasma cortisol levels and a Synacthen test may be indicated.
- ▶ For localized hyperpigmentation:
- Histopathologically may be indicated.
  - Imaging may be helpful whenever hyperpigmentation may be caused by amalgam, graphite or a foreign body.
  - Photographs may be useful for future comparison.
- Differential Diagnosis** ▶ Amalgam, graphite or other tattoos
- ▶ Ephelis (freckle)
  - ▶ Naevus
  - ▶ Melanotic macules
  - ▶ Melanoacanthoma
  - ▶ Malignant melanoma
  - ▶ Kaposi's sarcoma
  - ▶ Epithelioid angiomatosis
  - ▶ Verruciform xanthoma
  - ▶ Genetic:
    - Racial
    - Peutz-Jeghers syndrome
    - Laugier-Hunziker syndrome
    - Complex of myxomas, spotty pigmentation and endocrine overactivity
    - Carney syndrome



FIGURE 85

Gingival pigmentation:  
racial



FIGURE 86

Gingival pigmentation:  
racial



- Leopard syndrome
- Lentiginosis profusa
- ▶ Habits:
  - Using betel (brown)
  - Using the bark of *Juglans regia* for teeth cleaning (brown orange)
  - Chewing the seeds of *Cola nitida* (yellow)
  - Chewing the leaves of *Catha edulis* (brown)
- ▶ Drugs:
  - Smoking
  - Betel
  - Antimalarials
  - Amiodarone
  - Minocycline
  - Chlorpromazine
  - ACTH
  - Zidovudine
  - Clofazimine

- Ketoconazole
- Methyldopa
- Busulphan
- Menthol
- Contraceptive pill
- Metals
- ▶ Endocrine:
  - Addison's disease
  - Albright's syndrome
  - Pregnancy
- ▶ Post-inflammatory
- ▶ Others:
  - Hemochromatosis
  - Albright's syndrome
  - Generalized neurofibromatosis
  - Incontinentia pigmenti
  - Whipple's disease
  - Wilson's disease
  - Gaucher's disease
  - HIV disease
  - Thalassemia
  - Jaundice

**Treatment** ▶ Reassurance

### Melanotic Macule

**Definition** ▶ A melanotic macule is a pale brown macule less than 3 mm in diameter, with a poorly defined margin.

**Etiology** ▶ There is overproduction of melanin.

**Gingival Involvement** ▶ Very rare

**Other Sites of Involvement** ▶ Mainly seen on the lips

**Main Clinical Features** ▶ A pale brown macule less than 3 mm in diameter, with a poorly defined margin (**Fig. 87**)

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Histopathology in some instances.

▶ Imaging can be added, which may be helpful whenever hyperpigmentation may be caused by amalgam, graphite or a foreign body.

▶ Photographs may also be useful for future comparison.

**Differential Diagnosis** ▶ Racial pigmentation

▶ Localized irritation, e.g. smoking

▶ Amalgam tattoo

▶ Malignant melanoma

▶ Kaposi's sarcoma

▶ Drugs, e.g. antimalarials

▶ Addison's disease

**Treatment** ▶ Reassurance

FIGURE 87

Gingival pigmentation:  
melanotic macule



### Naevus

**Definition** ▶ Naevi are circumscribed developmental anomalies, usually brown, blue or black pigmented lesions.

**Etiology** ▶ Melanocytic naevi can appear or increase in size and colour with age, at adolescence and in pregnancy or after the administration of ACTH. There are several types:

- Lentigo, a brown circular macule due to increased melanocytes. Occasionally these are seen in hereditary syndromes such as NAME, LAMB and LEOPARD syndromes.
- Intramucosal naevus, consisting of a collection of naevus cells in the lamina propria without involvement of the epithelium, is most common (about 60%).
- Blue naevus, consisting of spindle cells at any level in the lamina propria, accounts for 25%.
- ▶ Others:
  - Junctional naevi, rare in the mouth.
  - Compound naevi, rare in the mouth.
  - Combined naevi, rare in the mouth.
  - Naevus of Ota, rare in the mouth.
  - Congenital naevi, rare in the mouth.
  - Dysplastic naevi have not been reported.

**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ Any oral site, particularly on the palate or buccal mucosa

**Main Clinical Features** ▶ Usually brown (**Figs. 88–92**)  
 ▶ Macular or papular  
 ▶ Do not change rapidly in size or colour  
 ▶ Painless



FIGURE 88

Gingival pigmentation:  
ephelis



FIGURE 89

Gingival pigmentation:  
compound naevus



FIGURE 90

Gingival pigmentation:  
naevus of Ota





FIGURE 91

Gingival pigmentation:  
naevus of Ota



FIGURE 92

Ocular pigmentation:  
naevus of Ota



- Diagnosis**
- ▶ Diagnosis is from history and clinical features, supplemented by:
    - Histopathology.
  - ▶ Imaging may be helpful whenever hyperpigmentation may be caused by amalgam, graphite or a foreign body.
  - ▶ Photographs may be useful for future comparison.
- Differential Diagnosis**
- ▶ Racial pigmentation
  - ▶ Localized irritation, e.g. smoking
  - ▶ Amalgam tattoo
  - ▶ Melanotic macule
  - ▶ Malignant melanoma
  - ▶ Kaposi's sarcoma
  - ▶ Drugs, e.g. antimalarials
  - ▶ Addison's disease
- Treatment**
- ▶ Excision is recommended both for cosmetic reasons and to exclude malignancy, especially melanoma.

### Metal Tattoo

- Definition** ▶ A hyperpigmented lesion caused by embedded metal particles or salts
- Etiology** ▶ Dental amalgam incorporated into the tissues during conservative dentistry or endodontics is the common cause.
- Gingival Involvement** ▶ Common
- Other Sites of Involvement** ▶ Typically in the mandibular area, close to the teeth
- Main Clinical Features** ▶ Bluish-black macule (**Figs. 93–97**), especially in the gingiva or vestibule in the premolar–molar region of the mandible
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:
- Histopathology..
- ▶ Imaging may be helpful whenever hyperpigmentation may be caused by amalgam, graphite or a foreign body.
- ▶ Photographs may be useful for future comparison.
- Differential Diagnosis** ▶ Racial pigmentation
- ▶ Localized irritation, e.g. smoking
- ▶ Naevus
- ▶ Melanotic macule
- ▶ Malignant melanoma
- ▶ Kaposi's sarcoma
- ▶ Drugs, e.g. antimalarials
- ▶ Addison's disease
- Treatment** ▶ Reassurance or, if in doubt, excise

**FIGURE 93**

Gingival pigmentation:  
amalgam tattoo



FIGURE 94

Gingival pigmentation:  
amalgam tattoo



FIGURE 95

Alveolar mucosa pigmentation:  
amalgam tattoo



FIGURE 96

Gingival pigmentation:  
tattoo from palladium crown





**FIGURE 97**

Gingival pigmentation:  
graphite tattoo

**FIGURE 98**

Gingival pigmentation:  
smoker's melanosis



### Smoker's Melanosis

**Definition** ▶ Hyperpigmentation induced by tobacco smoking

**Etiology** ▶ Tobacco smoking

**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ Buccal mucosa

**Main Clinical Features** ▶ Brown hyperpigmentation (**Fig. 98**)

**Diagnosis** ▶ Diagnosis is from smoking history and clinical features, supplemented by:

- Histopathology.
- ▶ Imaging may be helpful whenever hyperpigmentation may be caused by amalgam, graphite or a foreign body.
- ▶ Photographs may be useful for future comparison.



- Differential Diagnosis**
- ▶ Racial pigmentation
  - ▶ Amalgam tattoo
  - ▶ Naevus
  - ▶ Melanotic macule
  - ▶ Malignant melanoma
  - ▶ Kaposi's sarcoma
  - ▶ Drugs, e.g. antimalarials
  - ▶ Addison's disease
- Treatment**
- ▶ Reassurance
  - ▶ Stop habit

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# Allergic and Foreign Body Reactions

## Reactions to Dental Materials and Oral Health Care Products

- Definition** ▶ Reactions to:
- Materials and foods.
  - Deliberate tattoos.
- ▶ Fragments of teeth, dental materials, glass and other materials traumatically introduced into the tissues.
- ▶ Shrapnel and other materials can also be accidentally introduced.
- ▶ Material close to the gingiva may produce foreign body gingivitis.
- Etiology** ▶ Allergic reactions may be to oral health care products or foods or other antigens.
- ▶ Foreign bodies are introduced mainly in dental, sports or road traffic accidents, or assaults.
- ▶ Gingival inflammation associated with foreign bodies in the connective tissue is termed foreign body gingivitis (FBG). It is not commonly recognized by clinicians. It is more common in females. Most frequently, a red or red-and-white painful, chronic lesion, it has usually been present for less than 1 year and does not resolve with optimization of oral hygiene. It may be clinically confused with lichen planus. There is no gingival site predilection. Microscopically, foreign bodies are associated with the gingival inflammation, and elemental analysis suggests that they are usually derived from abrasives and less commonly from restorative materials.
- Gingival Involvement** ▶ Rare
- Other Sites of Involvement** ▶ Various
- Main Clinical Features** ▶ There may be pigmentation (**Fig. 99**), inflammation (**Figs. 100, 101**), whitish areas of necrotic epithelium (**Fig. 102**) or sometimes, no features.
- Diagnosis** ▶ Diagnosis is from history and clinical features. Wounds should always be examined by:
- Inspection.
  - Probing.
  - Imaging.
  - Ultrasonography (sometimes).
- Differential Diagnosis** ▶ Naevus
- ▶ Neoplasm

FIGURE 99

Gingival pigmentation:  
deliberate tattoo



FIGURE 100

Contact gingivitis from chlorhexidine



FIGURE 101

Contact gingivitis from Hexalen





**FIGURE 102**

Contact gingivitis from cinnamon



- Treatment**
- ▶ Laser or cryosurgery has been used to remove the foreign material.
  - ▶ Foreign material is best removed at an early stage as it may eventually form:
    - Tattoos.
    - Granulomas.
    - Scars.

### **Materia Alba**

- Definition** ▶ Debris consisting of squames, food, microorganisms and other material
- Etiology** ▶ Poor oral hygiene
- Gingival Involvement** ▶ Common
- Other Sites of Involvement** ▶ None
- Main Clinical Features** ▶ Whitish plaques that easily can be removed with gauze (**Figs. 103, 104**)
- Diagnosis** ▶ Diagnosis is from history and clinical features.
- Differential Diagnosis**
- ▶ Candidiasis
  - ▶ Lichen planus
  - ▶ Keratosis
- Treatment** ▶ Oral hygiene

FIGURE 103

Materia alba



FIGURE 104

Materia alba



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### Necrotizing Ulcerative Gingivitis

- ▶ Acute necrotizing ulcerative gingivitis (ANUG)
- ▶ Vincent's disease
- ▶ Trench mouth

**Definition** ▶ Non-contagious anaerobic infection associated with overwhelming proliferation of *Borrelia vincentii* and fusiform bacteria

**Etiology** ▶ Anaerobic flora implicated includes fusiform bacteria and spirochaete, with proliferation of *Borrelia vincentii*, *Prevotella intermedia*, fusiform bacilli, *Selenomonas* and other anaerobes.

▶ Predisposing factors include:

- Smoking.
- Viral respiratory infections.
- Poor oral hygiene.
- Malnutrition.
- Immune defects, including HIV and other viral infections, and leukaemias.

**Gingival Involvement** ▶ Uncommon except in:

- Lower socioeconomic classes
- HIV and other immune defects

**Other Sites of Involvement** ▶ Occasional spread to contiguous or adjacent tissues

**Main Clinical Features** ▶ Typically seen in adolescents and young adults, especially in institutions, armed forces, etc.

▶ Painful ulceration of the interdental papillae (**Figs. 105–108**).

▶ Pronounced tendency to gingival bleeding.

▶ Halitosis.

▶ Sialorrhoea.

▶ ANUG in malnourished, debilitated, or severely immunocompromised patients may extend onto the oral mucosa and skin, resulting in cancrum oris (noma; gangrenous stomatitis). Anaerobes, particularly bacteroides (*Porphyromonas*) species and *Fusobacterium necrophorum*, have been implicated and spreading necrosis eventually penetrates the buccal mucosa, leading to gangrene and ultimately a defect (oro-cutaneous fistula) and scarring.

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Smear for fusospirochaetal bacteria and leucocytes.
- Blood picture occasionally.



FIGURE 105

Necrotizing gingivitis:  
acute early



FIGURE 106

Necrotizing gingivitis:  
acute moderate



FIGURE 107

Necrotizing gingivitis:  
acute advanced





**FIGURE 108**

Necrotizing gingivitis:  
acute advanced



**Differential Diagnosis**

- ▶ Acute leukaemia
- ▶ Herpetic stomatitis

**Treatment**

- ▶ Oral debridement and hygiene instruction
- ▶ Peroxide or perborate mouthwashes
- ▶ Metronidazole
- ▶ Periodontal assessment

### Necrotizing Ulcerative Periodontitis

**Definition** ▶ Non-contagious anaerobic infection associated with overwhelming proliferation of *Borrelia vincentii* and fusiform bacteria

**Etiology** ▶ Predisposing factors include immune defects, particularly where there is also:

- Poor oral hygiene.
- Malnutrition.
- Smoking.
- Viral respiratory infections.

▶ Anaerobic flora implicated includes fusiform bacteria and spirochaete, with proliferation of *Borrelia vincentii*, *Prevotella intermedia*, fusiform bacilli, *Selenomonas* and other anaerobes.

**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ Occasional spread to contiguous or adjacent tissues (**Fig. 109**)

**Main Clinical Features**

- ▶ Painful ulceration
- ▶ Rapid periodontal destruction (**Fig. 110**).

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Smear for fusospirochaetal bacteria and leucocytes.
- Blood picture occasionally.

**FIGURE 109**

Necrotizing gingivitis:  
acute advanced with early noma

**FIGURE 110**

Necrotizing periodontitis



**Differential Diagnosis**

- ▶ Acute leukaemia
- ▶ Herpetic stomatitis

**Treatment**

- ▶ Oral debridement and hygiene instruction
- ▶ Peroxide or perborate mouthwashes
- ▶ Metronidazole
- ▶ Periodontal assessment

### Pericoronitis

**Definition** ▶ Inflammation of the operculum over an erupting or impacted tooth

**Etiology** ▶ Accumulation of plaque/debris beneath an operculum and irritation by an occluding tooth

**Gingival Involvement** ▶ Common in adolescents and teenagers

**FIGURE 111**

Pericoronitis  
(the white area is caused  
by materia alba)



**Other Sites of Involvement** ▶ None

- Main Clinical Features** ▶ The lower third molar is the site most commonly affected, but it is occasionally seen in relation to second molars. Patients complain of:
- Pain.
  - Trismus.
  - Swelling of operculum (**Fig. 111**).
  - Halitosis.
- ▶ In acute pericoronitis, there may also be:
- Swollen, red and often ulcerated operculum.
  - Pus from beneath operculum.
  - Some facial swelling.
  - Fever.
  - Regional lymphadenitis.
  - Malaise.

**Diagnosis** ▶ Diagnosis is from history and clinical features supplemented by imaging, which is necessary to define the prognosis for the tooth involved.

**Differential Diagnosis** ▶ ANUG  
▶ Leukaemia  
▶ Infectious mononucleosis

- Treatment** ▶ Remove any irritation from an opposing occluding tooth cusp.  
▶ Irrigate the operculum with saline.  
▶ Possibly apply astringent such as trichloroacetic acid to the operculum undersurface.  
▶ Antimicrobials such as metronidazole or penicillin if there are systemic symptoms or signs.  
▶ Consider removing the offending tooth after acute features have subsided.

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### Herpetic Gingivitis

#### Primary

- Definition** ▶ Primary infection with herpes simplex virus (HSV)
- Etiology** ▶ HSV-type 1 usually, transmitted in saliva  
▶ Rarely HSV-type 2
- Gingival Involvement** ▶ Very common; primary herpetic gingivostomatitis is the most common acute viral infection of oral mucosa  
▶ Non-plaque-induced lesions with no attachment loss
- Other Involvement** ▶ Tongue, buccal mucosa, palate, lips
- Main Clinical Features** ▶ Acute onset.  
▶ Erythema, gingival swelling.  
▶ Vesicles.  
▶ Then erosions (**Figs. 112–115**) and pain.  
▶ High fever, regional lymphadenopathy.  
▶ General malaise.  
▶ The lesions persist for 1–2 weeks and heal without scarring.
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:  
• HSV immunostaining.  
• Culture.
- Differential Diagnosis** ▶ Necrotizing ulcerative gingivitis  
▶ Desquamative gingivitis  
▶ Erythema multiforme  
▶ Pemphigus vulgaris
- Treatment** ▶ Symptomatic  
▶ Aciclovir systemically as early as possible

FIGURE 112

Infective gingivitis:  
primary herpes simplex stomatitis



FIGURE 113

Infective gingivitis:  
primary herpes simplex stomatitis



FIGURE 114

Infective gingivitis:  
primary herpes simplex stomatitis



**FIGURE 115**

Infective gingivitis:  
primary herpes simplex stomatitis



### Secondary

**Definition** ▶ Recurrent infection with herpes simplex virus (HSV)

**Etiology** ▶ HSV, usually type 1  
 ▶ Most likely:

- After trauma, such as a local analgesic injection
- In immunosuppressed patient, e.g. HIV or leukaemia

**Gingival Involvement** ▶ Uncommon

**Other Sites of Involvement** ▶ Any oral site

**Main Clinical Features** ▶ Multiple vesicles and round scattered ulcers with yellow slough and erythematous halo (**Figs. 116–118**).  
 ▶ Ulcers fuse to produce irregular lesions, typically in palate or gingiva.

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Serology, which is confirmatory but rarely required. A rising titre of antibodies is confirmatory.
- ▶ Smear for viral damaged cells, viral culture or electron microscopy is used occasionally.

**Differential Diagnosis** ▶ Other causes of mouth ulcers, especially:

- Hand, foot and mouth disease
- Herpangina
- Shingles
- Erythema multiforme
- Leukaemia

**Treatment** ▶ Soft diet  
 ▶ Adequate fluid intake  
 ▶ Antipyretics/analgesics (paracetamol elixir)  
 ▶ Local antiseptics (0.2% aqueous chlorhexidine mouthwashes)  
 ▶ Aciclovir orally or parenterally in immunocompromised patients



FIGURE 116

Infective gingivitis:  
recurrent herpes simplex stomatitis



FIGURE 117

Infective gingivitis:  
recurrent herpes simplex stomatitis



FIGURE 118

Infective gingivitis:  
recurrent herpes simplex stomatitis





**FIGURE 119**

Infective gingivitis:  
human papillomavirus;  
condyloma acuminatum



## Papillomaviruses

### Condyloma Acuminatum

- Definition** ▶ Infection with human papillomavirus (HPV)
- Etiology** ▶ HPV-related lesion transmitted by orogenital or oroanal sex
- Gingival Involvement** ▶ Rare
- Other Sites of Involvement** ▶ Anogenital region
- Main Clinical Features** ▶ Warty papules or more smooth-surfaced lesions (**Fig. 119**)  
▶ Occasionally on tongue or palate
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:  
• Histopathology.
- Differential Diagnosis** ▶ Other HPV-related lesions such as papilloma
- Treatment** ▶ Medical:  
• Podophyllum  
• Imiquimod  
• Intralesional interferon alpha  
▶ Surgical:  
• Excision  
• Laser  
• Electrosurgery  
• Cryosurgery

### Verruca Vulgaris

- Definition** ▶ Infection with HPV
- Etiology** ▶ Infection with HPV from a skin wart

FIGURE 120

Infective gingivitis:  
human papillomavirus;  
verruca vulgaris



FIGURE 121

Infective gingivitis:  
human papillomavirus;  
verruca vulgaris



FIGURE 122

Infective gingivitis:  
human papillomavirus;  
verruca vulgaris



- Gingival Involvement** ▶ Rare
- Other Sites of Involvement** ▶ Mainly skin
- Main Clinical Features** ▶ Warty papules or more smooth-surfaced lesions (**Figs. 120–122**)
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:
- Histopathology to differentiate from other HPV-related lesions and tumours.
- Differential Diagnosis** ▶ Other HPV-related lesions such as papilloma
- Treatment** ▶ Medical:
- Podophyllum
  - Imiquimod
  - Intralesional interferon alpha
- ▶ Surgical:
- Excision
  - Laser
  - Electrosurgery
  - Cryosurgery

### Papilloma

- Definition** ▶ Infection with HPV, usually HPV-6 or 11
- Etiology** ▶ HPV infections
- Gingival Involvement** ▶ Uncommon
- ▶ The most common benign soft tissue neoplasm
  - ▶ Found in the 20–50-year age group
- Other Sites of Involvement** ▶ Mainly lip, sometimes palate, tongue or other sites
- Main Clinical Features** ▶ Typically asymptomatic
- ▶ Warty papules or more smooth-surfaced lesion (**Figs. 123, 124**)
  - ▶ Pedunculated lesion, either pink or white if hyperkeratinized
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:
- Histopathology.
- Differential Diagnosis** ▶ Other HPV-related lesions such as condyloma
- Treatment** ▶ Medical:
- Podophyllum
  - Imiquimod
  - Intralesional interferon alpha
- ▶ Surgical:
- Excision
  - Laser
  - Electrosurgery
  - Cryosurgery

FIGURE 123

Infective gingivitis:  
human papillomavirus;  
papilloma



FIGURE 124

Infective gingivitis:  
human papillomavirus;  
papilloma





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## Fungal Infections

### Acute Pseudomembranous Candidiasis

**Definition** ► Infection with yeasts of the species *Candida*

**Etiology** ► Predisposing factors to pseudomembranous candidiasis include:

- *C. albicans*.
- *C. tropicalis*.
- *C. glabrata*.
- *C. parapsilosis*.
- *C. krusei*.
- Other *Candida* species.
- Other genera (*Rhodotorula*, *Saccharomyces*, etc.), which are rare and transient.

► Factors that can increase the liability to candidiasis include:

- Xerostomia.
- Smoking.
- Use of corticosteroids systemically or in the mouth/pharynx.
- Use of broad-spectrum antimicrobials.
- Cytotoxic chemotherapy.
- Irradiation involving the mouth/salivary glands.
- Malnutrition.
- Appliances.
- Immune defects. Candidiasis may be seen in healthy neonates (who have immature immune systems and no acquired immunity). Any T cell defect, especially HIV infection, immunosuppressive and cytotoxic treatment, leukaemias and lymphomas, and cancer can predispose to candidiasis and other fungal infections. Candidiasis can also be seen where there are neutrophil defects such as in diabetes.

**Gingival Involvement** ► Common

**Other Sites of Involvement** ► Any oral or mucocutaneous site

**Main Clinical Features** ► White or creamy plaques that can be wiped off to leave a red base (Fig. 125)

**Diagnosis** ► Diagnosis is from history and clinical features, supplemented by:

- Gram-stained smear (hyphae) or oral rinse.

**Differential Diagnosis** ► Koplik's spots  
► Fordyce's spots  
► Lichen planus

FIGURE 125

Infective gingivitis:  
candidiasis; pseudomembranous



- Treatment**
- ▶ Avoid or reduce smoking.
  - ▶ Treat any local predisposing cause such as xerostomia.
  - ▶ Improve oral hygiene.
  - ▶ Antifungals such as nystatin oral suspension or pastilles or amphotericin lozenges or miconazole gel or tablets or fluconazole tablets or suspension. Itraconazole in the immunocompromised.
  - ▶ Chlorhexidine and triclosan have some anti-candidal activity.

### Erythematous Candidiasis

- Definition** ▶ Red lesions due to *Candida*

- Etiology**
- ▶ Predisposing factors to erythematous candidiasis include:
    - *C. albicans*.
    - *C. tropicalis*.
    - *C. glabrata*.
    - *C. parapsilosis*.
    - *C. krusei*.
    - Other *Candida* species.
    - Other genera (*Rhodotorula*, *Saccharomyces*, etc.), which are rare and transient.
  - ▶ Red lesions may be seen in denture-induced stomatitis, antibiotic-induced stomatitis, sometimes in median rhomboid glossitis, and in HIV infection may develop de novo, may precede pseudomembranous candidiasis, or may arise as a consequence of persistent acute pseudomembranous candidiasis when the pseudomembranes are shed. Factors that can increase the liability to candidiasis include:
    - Xerostomia.
    - Smoking.
    - Use of corticosteroids systemically or in the mouth/pharynx.
    - Use of broad-spectrum antimicrobials.
    - Cytotoxic chemotherapy.
    - Irradiation involving the mouth/salivary glands.
    - Malnutrition.
    - Appliances.



FIGURE 126

Infective gingivitis:  
candidiasis; erythematous



FIGURE 127

Infective gingivitis:  
candidiasis; erythematous,  
denture-induced



- Immune defects. Candidiasis may be seen in healthy neonates (who have immature immune systems and no acquired immunity). Any T cell defect, especially HIV infection, immunosuppressive and cytotoxic treatment, leukaemias and lymphomas, and cancer can predispose to candidiasis and other fungal infections. Candidiasis can also be seen where there are neutrophil defects such as in diabetes.

**Gingival Involvement** ► Rare

- Other Sites of Involvement** ► Red lesions that involve the gingivae (Figs. 126, 127) may be seen in denture-induced stomatitis and antibiotic-induced stomatitis. Denture-induced stomatitis is usually chronic atrophic candidiasis and consists of mild inflammation of the mucosa beneath a denture – usually a complete upper denture. In HIV infection, erythematous candidiasis may develop de novo, may precede pseudomembranous candidiasis, or may arise as a consequence of persistent acute pseudomembranous candidiasis when the pseudomembranes are shed.



**Main Clinical Features** ▶ Erythema and soreness

**Diagnosis** ▶ This is a clinical diagnosis but if the etiology is not absolutely clear, a blood picture, and smears for fungal hyphae and culture may be helpful to management.

**Differential Diagnosis** ▶ Inflammatory gingivitis  
 ▶ Desquamative gingivitis  
 ▶ Trauma  
 ▶ Drugs and plasma cell gingivostomatitis  
 ▶ Infections, e.g. primary herpetic stomatitis, candidiasis  
 ▶ Granulomas, e.g. Crohn's disease, orofacial granulomatosis or sarcoidosis  
 ▶ Kaposi's sarcoma

**Treatment** ▶ Avoid or reduce smoking.  
 ▶ Treat any local predisposing cause such as xerostomia.  
 ▶ Improve oral hygiene.  
 ▶ Antifungals such as nystatin oral suspension *or* pastilles *or* amphotericin lozenges *or* miconazole gel *or* tablets *or* fluconazole tablets *or* suspension.  
 ▶ Chlorhexidine and triclosan have some anti-candidal activity.

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## Aphthous Ulceration

- Definition** ▶ Recurrent idiopathic mouth ulcers with onset usually in childhood or adolescence
- Etiology** ▶ Unclear: immunological changes are detectable but there is no reliable evidence of autoimmune disease or any classic immunological reactions. It may be due to:
- Changes in cell-mediated immune responses.
  - Cross-reactivity with *Streptococcus sanguis* or heat shock protein.
- ▶ Underlying predisposing factors seen in a minority include:
- Haematinic (iron, folate or vitamin B12) deficiency in 10 %–20 %.
  - Relationship with luteal phase of menstruation.
  - Stress (and stopping smoking).
  - Food allergies.
  - HIV disease.
- Gingival Involvement** ▶ Uncommon
- Other Sites of Involvement** ▶ None
- Main Clinical Features** ▶ Affects about 25 % of population, mostly non-smokers.
- ▶ Onset is usually in childhood or adolescence.
- ▶ Later onset may signify haematinic deficiency or HIV disease.
- ▶ Recurrent round or ovoid ulcers (**Figs. 128–132**) usually lasting 1 week or 1 month.
- ▶ There are three distinct clinical patterns of aphthae:
- Minor – small ulcers (< 4 mm) on mobile mucosae, healing within 14 days, no scarring (**Figs. 128–130**).
  - Major – large ulcers (may be > 1 cm), any site including dorsum of tongue and hard palate, healing within 1–3 months, with scarring (**Figs. 131, 132**).
  - Herpetiform ulcers – multiple minute ulcers that coalesce to produce ragged ulcers.
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:
- A blood picture, which is useful to reveal possible deficiencies.
- Differential Diagnosis** ▶ Local causes of ulcers
- ▶ Neoplasms
- ▶ PFAPA syndrome (periodic fever, aphthous stomatitis, pharyngitis, and adenopathy)
- ▶ Behçet's syndrome
- ▶ Systemic disease
- Infections:
  - Haematological disease

- Gastrointestinal disease
- Mucocutaneous disease
- Drugs
- Irradiation

- Treatment**
- ▶ Treat any underlying predisposing factors.
  - ▶ Treat aphthae with:
    - Chlorhexidine 0.2% aqueous mouthwash or tetracycline rinses.
    - Topical corticosteroids (hydrocortisone hemisuccinate 2.5 mg pellets or 0.1% triamcinolone acetonide in Orabase).
    - Sometimes more potent topical steroids (e.g. betamethasone, beclomethasone, fluocinonide or fluticasone).
    - Rarely, other agents such as levamisole or thalidomide.

FIGURE 128

Aphthous stomatitis:  
minor aphthae



FIGURE 129

Aphthous stomatitis:  
minor aphthae





**FIGURE 130**

Aphthous stomatitis:  
minor aphthae

**FIGURE 131**

Aphthous stomatitis:  
major aphthae

**FIGURE 132**

Aphthous stomatitis:  
major aphthae





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### Hereditary Gingival Fibromatosis

- Definition** ▶ Hereditary gingival fibromatosis is a rare genetic disorder that may exist as an isolated disease entity or as part of a syndrome.
- Etiology** ▶ Genetic. As a single disease entity it may be sporadic or may be transmitted as an autosomal dominant or rarely autosomal recessive trait.
- Gingival Involvement** ▶ Always present  
▶ Non-plaque-induced gingival lesions
- Other Sites of Involvement** ▶ May vary according to the syndrome it is associated with. No other involvement in isolated cases.
- Main Clinical Features** ▶ Severe generalized gingival overgrowth covering the teeth partially or completely is the characteristic feature (**Fig. 133, 134**).  
▶ Gingiva are smooth, of a firm consistency and usually normal in colour.  
▶ Appear during childhood and occasionally delay eruption of teeth.
- Diagnosis** ▶ Diagnosis is made from history and clinical features, supplemented by:  
• Biopsy and histopathological examination.
- Differential Diagnosis** ▶ Several syndromes associated with gingival fibromatosis (Laband, Cross, Ramon, Rutherford)  
▶ Drug-induced gingival overgrowth (phenytoin, ciclosporin, calcium channel blockers)  
▶ Familial acanthosis nigricans  
▶ Crohn's disease  
▶ Amyloidosis  
▶ Leukaemia
- Treatment** ▶ Gingivectomy  
▶ Dental plaque control

FIGURE 133

Congenital gingival fibromatosis.  
Generalized gingival overgrowth  
covering the teeth



FIGURE 134

Congenital gingival fibromatosis.  
Localized early gingival overgrowth



### White Sponge Naevus

**Definition** ▶ White sponge naevus, or Cannon's disease, is a relatively uncommon genetic disorder that primarily affects the oral mucosa.

**Etiology** ▶ Genetic. It is inherited as an autosomal dominant trait.

**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ Oral mucosa (buccal, tongue, floor of the mouth, labial), always present  
▶ Other mucosae (vaginal, rectal, oesophageal, nasal), rare

**Main Clinical Features** ▶ The gingival lesions present as painless white or grey-white, gingival and alveolar mucosa thickenings with multiple furrows, diffuse plaques with a spongy texture (**Fig. 135**).  
▶ Similar lesions appear more often in other sites of the oral mucosa.  
▶ Oral lesions become fully fledged by the second decade of life and remain relatively stable thereafter.



**FIGURE 135**

White sponge naevus.  
White thickening of the alveolar  
mucosa extending to the gingiva



- Diagnosis**
- ▶ The diagnosis is based on clinical and laboratory criteria.
  - ▶ Biopsy and histopathological examination.
  - ▶ Cytology is useful.
- Differential Diagnosis**
- ▶ Leukoplakia
  - ▶ Focal palmoplantar and oral mucosa hyperkeratosis syndrome
- Treatment**
- ▶ No treatment available
  - ▶ Surgical excision of gingival lesions only for aesthetic purposes

### Laband Syndrome

- Definition**
- ▶ Laband syndrome is a rare disorder characterized by a broad spectrum of lesions.
- Etiology**
- ▶ Genetic. It is inherited as an autosomal dominant trait.
- Gingival Involvement**
- ▶ Always present
  - ▶ Non-dental plaque-induced gingival lesions with no attachment loss
- Other Sites of Involvement**
- ▶ Oral mucosa, rare
  - ▶ Skin, facies, skeletal
  - ▶ Liver, spleen
- Main Clinical Features**
- ▶ Gingival fibromatosis is a constant feature and it is manifested at birth or within the first 2 months of life. The gingiva are characteristically enlarged and cover the crowns of the teeth partly or completely (**Fig. 136**). The enlarged gingiva are usually lobulated and slightly firm on palpation.
  - ▶ Macroglossia and enlargement of the lips are less common manifestations.
  - ▶ Large nose and pinnae, hypoplastic or absent thumb nails, rudimentary or missing terminal phalanges, pes cavus, joint hypermobility and hirsutism, are common (**Fig. 137**).
  - ▶ Hepatomegaly, splenomegaly and learning disability have been noted.

FIGURE 136

Laband syndrome.  
Gingival overgrowth covering  
the crowns of the teeth



FIGURE 137

Laband syndrome.  
Hypoplastic thumb nails



**Diagnosis** ► The diagnosis is based on clinical criteria.

**Differential Diagnosis** ► Hereditary gingival fibromatosis  
► Drug-influenced gingival enlargement (phenytoin, ciclosporin, calcium channel blockers)  
► Several syndromes associated with gingival fibromatosis (Rutherford syndrome, Murray-Puretic-Drescher syndrome, Cross and Ramon syndromes)  
► Alpha-mannosidosis

**Treatment** ► Good oral hygiene  
► Gingivectomy

### Papillon-Lefèvre Syndrome

- Definition** ▶ Papillon-Lefèvre is a rare genetic disorder characterized by periodontal and skin manifestations.
- Etiology** ▶ Genetic. It is inherited as an autosomal recessive trait.  
 ▶ Periodontal lesions are due to multiple leukocyte dysfunctions, and a specific bacterial infection of dental plaque origin (*Actinobacillus actinomycetemcomitans*).
- Periodontal Involvement** ▶ Always present  
 ▶ Periodontitis associated with genetic disorder
- Other Sites of Involvement** ▶ Skin, always present (palms, soles, elbows, knees, dorsum of the fingers and toes)
- Main Clinical Features** ▶ Clinically, the gingiva are red, swollen and bleed easily. Severe generalized periodontitis with deep periodontal pockets and bone loss are constant features (Fig. 138). Mobility, migration and early loss both of deciduous and permanent teeth following the order of eruption is the rule.  
 ▶ The skin lesions are characterized by hyperkeratosis of palms and soles (Figs. 139, 140). Hyperkeratosis and psoriasiform scaly red patches may occur in other skin areas.
- Diagnosis** ▶ The diagnosis is based on clinical criteria.  
 ▶ Panoramic radiography.  
 ▶ Skin biopsy and histopathological examination.
- Differential Diagnosis** ▶ Generalized aggressive periodontitis  
 ▶ Hypophosphatasia, acatalasia  
 ▶ Chediak-Higashi syndrome  
 ▶ Langerhans cell histiocytosis  
 ▶ Glycogen storage disease 1b  
 ▶ Congenital neutropenia, cyclic neutropenia
- Treatment** ▶ Dental plaque control  
 ▶ Good oral hygiene  
 ▶ Aromatic retinoids for skin lesions



FIGURE 138

Papillon-Lefèvre syndrome.  
Severe generalized periodontitis  
in a 13-year-old boy



FIGURE 139

Papillon-Lefèvre syndrome.  
Hyperkeratosis of the palms



FIGURE 140

Papillon-Lefèvre syndrome.  
Hyperkeratosis of the soles





## Hypophosphatasia

- Definition** ▶ Hypophosphatasia is a rare inherited enzyme deficiency disorder, characterized by decreased serum and tissue alkaline phosphate production and activity.
- Etiology** ▶ Genetic. It is usually inherited as an autosomal recessive trait and rarely as an autosomal dominant trait.
- Periodontal Involvement** ▶ Very common, particularly in the childhood form of the disease  
▶ Periodontitis associated with genetic disorder
- Other Sites of Involvement** ▶ Skeleton, skull, eyes
- Main Clinical Features** ▶ The periodontal manifestations include severe loss of alveolar bone and premature loss of deciduous and permanent teeth in the absence of an inflammatory response (**Fig. 141**). Early exfoliation particularly affects the anterior teeth and may be the only clinical sign of the disease.  
▶ Skeletal abnormalities, growth retardation, craniosynostosis, intracranial hypertension may occur.
- Diagnosis** ▶ Diagnosis is made from history and clinical features, supplemented by:  
• Panoramic radiography.  
• Radiographic examination of the skeleton and skull.  
• Measurement of serum alkaline phosphatase and blood and urine phosphoethanolamine.
- Differential Diagnosis** ▶ Localized aggressive periodontitis  
▶ Acatalasia  
▶ Papillon-Lefèvre syndrome  
▶ Chediak-Higashi syndrome  
▶ Glycogen storage disease 1b
- Treatment** ▶ Good oral hygiene.  
▶ Vitamin D, phosphorus and parathormone have been used with poor results.

**FIGURE 141**

Hypophosphatasia.  
Premature loss of deciduous tooth  
and absence of inflammatory  
response



FIGURE 142

Glycogen storage disease, type 1b.  
Localized early onset gingivitis



FIGURE 143

Glycogen storage disease, type 1b.  
Generalized severe periodontitis



### Glycogen Storage Disease 1b

- Definition** ▶ Glycogen storage disease 1b is a rare metabolic disorder with severe prognosis.
- Etiology** ▶ Genetic. It is inherited as an autosomal recessive trait.  
▶ This is a defect of the microsomal glucose-6-phosphate translocase. The disorder is associated with neutropenia and neutrophil dysfunction.
- Periodontal Involvement** ▶ Relatively common  
▶ Periodontitis associated with genetic disorder
- Other Sites of Involvement** ▶ Oral mucosa, frequent  
▶ Liver, kidneys, skeleton and many other organs

**FIGURE 144**

Glycogen storage disease, type 1b.  
Characteristic “doll’s” face



- Main Clinical Features**
- ▶ Early onset chronic gingivitis soon leading to severe periodontal disease with alveolar bone loss and teeth migration (**Figs. 142, 143**).
  - ▶ The oral mucosal lesions present as discrete, deep, irregular, recurrent ulcers that are usually covered by a whitish pseudomembrane. Vaulted palate and recurrent oral infection may occur.
  - ▶ Characteristic face (doll's face), liver and kidney enlargement, hypoglycaemia, hyperlipidaemia, growth retardation and recurrent bacterial infection are common (**Fig. 144**).

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Haematological and biochemical examination.
  - ▶ Liver biopsy and histopathological examination.

- Differential Diagnosis**
- ▶ Congenital neutropenia
  - ▶ Cyclic neutropenia
  - ▶ Chediak-Higashi syndrome
  - ▶ Hypophosphatasia and acatalasia

- Treatment**
- ▶ Dental plaque control.
  - ▶ Topical oral antiseptics.
  - ▶ Systemic treatment by the paediatrician.

### Hurler's Syndrome

- Definition**
- ▶ Hurler's syndrome is a rare, and the most common and severe form, in the group of mucopolysaccharide metabolic disorders.

- Etiology**
- ▶ Genetic. It is inherited as an autosomal recessive trait.
  - ▶ The basic defect is the deficiency of the enzyme alpha-L-iduronidase resulting in the accumulation and deposition of heparan sulphate and dermatan sulphate within the tissues and their excessive excretion in the urine.

- Gingival Involvement**
- ▶ Common



FIGURE 145

Hurler's syndrome.  
Severe gingival overgrowth  
completely covering the crowns  
of the teeth

**Other Sites of Involvement**

- ▶ Oral mucosa and teeth
- ▶ Skeletal
- ▶ Cornea
- ▶ Internal organ (liver, spleen, heart, lungs and others)

**Main Clinical Feature**

- ▶ Gingival overgrowth that may partially or fully cover the crowns of teeth may be present, particularly in the anterior area of the maxilla (**Fig. 145**). The overgrowth is probably due to mouth breathing in association with microbial dental plaque and the deposition of heparan and dermatan sulphate within the gingiva.
- ▶ Other oral manifestations include macrocheilia, macroglossia, open mouth with protruding tongue, numerous impacted teeth, large interdental spaces and teeth dislocation.
- ▶ Growth retardation, craniofacial malformations, characteristic facies, scoliosis, stiff joints and chondrodystrophy are common.
- ▶ Corneal clouding that frequently leads to blindness, learning disability, cardiac failure, hypertension, lung infection may also occur.
- ▶ The disease presents during infancy and often leads to death usually before the patient reaches 10 years of age.

**Diagnosis**

- ▶ The diagnosis is based on clinical and laboratory criteria:
  - Histochemical evaluation of alpha-L-iduronidase.
  - Detection of heparan sulphate and dermatan sulphate in the urine.
  - Radiographic examination.

**Differential Diagnosis**

- ▶ Hereditary gingival fibromatosis
- ▶ Mouth-breathing gingivitis
- ▶ Other forms of mucopolysaccharidoses
- ▶ Alfa-mannosidosis

**Treatment**

- ▶ Good oral hygiene and gingivectomy
- ▶ Management of dental problems
- ▶ Supportive for the other manifestations



### Focal Palmoplantar and Oral Mucosa Hyperkeratosis Syndrome

- Definition** ▶ Focal palmoplantar and oral mucosa hyperkeratosis syndrome, or hyperkeratosis palmoplantaris and attached gingival hyperkeratosis is an unusual genetic disorder characterized by oral mucosal and dermal hyperkeratosis.
- Etiology** ▶ Genetic. It is inherited as an autosomal dominant trait.
- Gingival Involvement** ▶ Always present, mainly attached gingiva
- Other Sites of Involvement** ▶ Oral mucosa (sites subject to mechanical pressure or friction such as hard palate, retromolar pad mucosa, alveolar mucosa, lateral of the tongue)  
▶ Skin (mainly over the weight-bearing and pressure-related areas, such as the soles and palms)  
▶ Nails
- Main Clinical Features** ▶ Marked white hyperkeratosis of the attached gingiva, presenting clinically as leukoplakia, is a consistent finding (**Fig. 146**). There is no inflammation or attachment loss.  
▶ White hyperkeratotic lesions in other mucosal sites are less frequently observed.  
▶ White hyperkeratotic lesions at the weight-bearing and pressure-related areas of the palms and soles are always present (**Figs. 147, 148**).  
▶ Nail dystrophy and hyperhidrosis may also occur.  
▶ The hyperkeratosis develops early in childhood and the severity increases with age.
- Diagnosis** ▶ The diagnosis is mainly based on clinical criteria.  
▶ Biopsy and histopathological examination are also used.
- Differential Diagnosis** ▶ Leukoplakia  
▶ White sponge naevus  
▶ Pachyonychia congenita  
▶ Dyskeratosis congenita
- Treatment** ▶ Local and systemic aromatic retinoids with partial success

FIGURE 146

Focal palmoplantar and oral mucosa hyperkeratosis syndrome.  
Generalized white hyperkeratosis of the gingiva



FIGURE 147

Focal palmoplantar and oral mucosa hyperkeratosis syndrome.  
Hyperkeratosis of the palms



FIGURE 148

Focal palmoplantar and oral mucosa hyperkeratosis syndrome.  
Hyperkeratosis of the sole



### Familial Acanthosis Nigricans

- Definition** ▶ Familial or juvenile acanthosis nigricans is an uncommon benign mucocutaneous disorder, characterized by papillary lesions and occasional skin discolouration.
- Etiology** ▶ Genetic. It is inherited by an irregular autosomal dominant trait.
- Gingival Involvement** ▶ Relatively common  
▶ Non-dental plaque-related lesions
- Other Sites of Involvement** ▶ Oral mucosa (10 %–25 %), mainly on the tongue, palate, lips  
▶ Skin, always (axillae, groins, neck, umbilicus, genitalia, perianal region)
- Main Clinical Features** ▶ Gingival enlargement and in particular the interdental papillae may be present. The enlargement is occasionally so severe that it covers the crowns of teeth. Enlarged gingiva often end up looking papillary in appearance (**Figs. 149, 150**).  
▶ The oral mucosal lesions appear as multiple, small, painless papillomatous growths. The gingival and oral mucosal lesions exhibit normal colour.  
▶ The skin lesions present as painless dark, velvety, multiple small papillary growths (skin tags) (**Fig. 151**).  
▶ The disorder usually develops during childhood or at puberty.
- Diagnosis** ▶ The diagnosis is usually based on clinical criteria.  
▶ Biopsy and histopathological examination of oral and skin lesions are supportive.
- Differential Diagnosis** ▶ Hereditary gingival fibromatosis  
▶ Drug-induced gingival hyperplasia  
▶ Malignant acanthosis nigricans  
▶ Endocrine-related acanthosis nigricans  
▶ Cowden's disease  
▶ Darier's disease
- Treatment** ▶ Gingivectomy and good oral hygiene for the gingival lesions  
▶ Surgical reconstruction of skin lesion in cases of severe aesthetic problems



FIGURE 149

Familial acanthosis nigricans.  
Localized gingival enlargement  
with a papillary pattern



FIGURE 150

Familial acanthosis nigricans.  
Generalized papillary overgrowth  
of the alveolar mucosa



FIGURE 151

Familial acanthosis nigricans.  
Multiple small papillary growths  
of the skin (skin tags)





### Darier's Disease

- Definition** ▶ Darier's disease or dyskeratosis follicularis is a relatively rare mucocutaneous disorder.
- Etiology** ▶ Genetic. It is inherited as an autosomal dominant trait.
- Gingival Involvement** ▶ Relatively common  
▶ Non-dental plaque-induced gingival lesions
- Other Sites of Involvement** ▶ Oral mucosa (20%–40%)  
▶ Other mucosae (genitalia, rectum, pharynx)  
▶ Skin (always), nails
- Main Clinical Features** ▶ The gingival lesions appear as small multiple, whitish confluent papules which may become hypertrophic and are arranged in a reticular pattern or are assuming a cobblestone appearance (**Fig. 152**).  
▶ Similar lesions appear on the other oral mucosa area, particularly on the palate, tongue and buccal mucosa. The oral lesions usually appear after the cutaneous ones.  
▶ The skin lesions appear as multiple brownish-red, painless, papules that usually coalesce into plaques (**Fig. 153**). The forehead, ears, scalp, chest and back are more often affected.  
▶ The nails exhibit subungual keratosis and longitudinal ridges and lines.
- Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.  
▶ Biopsy and histopathological examination of oral and skin lesions.
- Differential Diagnosis** ▶ Acanthosis nigricans  
▶ Cowden's disease  
▶ Wegener's granulomatosis  
▶ Lichen planus
- Treatment** ▶ Good oral hygiene  
▶ Systemic aromatic retinoids

**FIGURE 152**

Darier's disease.  
Multiple, small whitish papules  
on the lower gingiva



FIGURE 153

Darier's disease.  
Multiple papules that coalesce  
into plaques on the skin



### Cowden's Disease

**Definition** ▶ Cowden's disease or multiple hamartoma syndrome is an uncommon disorder characterized by a wide spectrum of mucocutaneous lesions usually associated with breast, thyroid, and other organ disorders.

**Etiology** ▶ Genetic. It is inherited as an autosomal dominant trait.

**Gingival Involvement** ▶ Relatively common  
▶ Non-dental plaque-induced gingival lesions

**Other Sites of Involvement** ▶ Oral mucosa, common (lips, tongue, buccal mucosa)  
▶ Skin, common (neck, nasolabial folds, dorsum of the hands and forearms)  
▶ Breast, thyroid gland, common  
▶ Other organs (genitourinary system, gastrointestinal tract, skeletal, eye)

**Main Clinical Features** ▶ The gingival lesions appear as multiple, small, smooth, pale or whitish, painless papules or nodules that may be isolated or coalesce in a cobblestone pattern on the free and attached gingiva or the edentulous alveolar mucosa (Figs. 154, 155).  
▶ The oral lesions are similar to those seen on the gingiva.  
▶ The skin lesions appear as multiple papillomatous papules and small nodules that are hair follicle hamartomas (trichilemmomas). Dermal fibromas are also common.  
▶ Several breast and thyroid glands abnormalities including benign and malignant tumours are common.

**Diagnosis** ▶ The diagnosis is based on clinical and laboratory criteria.  
▶ Biopsy and histopathological examination is mainly of cutaneous lesions.

**FIGURE 154**

Cowden's disease.  
Multiple, small nodules of the gingiva

**FIGURE 155**

Cowden's disease.  
Multiple nodules that coalesce in a  
cobblestone pattern on the gingiva  
and alveolar mucosa

**Differential Diagnosis**

- ▶ Focal epithelial hyperplasia
- ▶ Acanthosis nigricans
- ▶ Tuberous sclerosis
- ▶ Multiple endocrine neoplasia syndrome
- ▶ Darier's disease
- ▶ Lipoid proteinosis

**Treatment**

- ▶ There is no treatment
- ▶ Surgical excision of oral and cutaneous lesions only for aesthetic purposes



## Tuberous Sclerosis

**Definition** ▶ Tuberous sclerosis of Bourneville-Pringle syndrome, or epiloia, is an uncommon disorder characterized by mucocutaneous angiofibromas, learning disability and seizures.

**Etiology** ▶ Genetic. It is inherited as an autosomal dominant trait, with variable expressivity.

**Gingival Involvement** ▶ Relatively common (10 %–15 %)  
▶ Non-dental plaque-induced gingival lesions

**Other Sites of Involvement** ▶ Oral mucosa, rare  
▶ Skin, always  
▶ Central nervous system  
▶ Others (retinal, heart, kidneys, lungs)

**Main Clinical Features** ▶ The gingival lesions are the most characteristic oral lesions and present as multiple, painless, fibrous nodules with smooth surface and normal or whitish colour (**Figs. 156, 157**). The anterior gingiva is more frequently affected.  
▶ The oral mucosal lesions, although less often, are similar to those seen on the gingiva. Cleft palate and lip, vaulted palate, macroglossia and enamel pits may occur.  
▶ The skin lesions are common and present as multiple small, red to brown nodules (angiofibromas), mainly in the nasolabial folds, and elsewhere in the face (**Fig. 158**). Periungual fibromas may also seen.  
▶ Learning disability, seizures, and neural hamartomas are common.  
▶ Cardiac rhabdomyoma, multiple retinal hamartomata, lung and renal abnormalities may also occur.

**Diagnosis** ▶ The diagnosis is based on clinical and laboratory criteria.  
▶ Biopsy and histopathological examination of cutaneous and oral lesions.  
▶ Brain CT scan.  
▶ Electroencephalography.

**Differential Diagnosis** ▶ Cowden's disease  
▶ Neurofibromatosis  
▶ Lipoid proteinosis  
▶ Acanthosis nigricans  
▶ Focal dermal hypoplasia syndrome

**Treatment** ▶ Symptomatic  
▶ Surgical excision of gingival lesions only for aesthetic purposes



**FIGURE 156**

Tuberous sclerosis.  
Solitary nodule on the gingiva

**FIGURE 157**

Tuberous sclerosis.  
Multiple gingival nodules

**FIGURE 158**

Tuberous sclerosis.  
Characteristic angiofibromas  
on the skin of the face



### Gardner's Syndrome

- Definition** ▶ Gardner's syndrome is a rare disorder in the group of familial colorectal polyposis syndromes.
- Etiology** ▶ Genetic. It is inherited as an autosomal dominant trait.
- Alveolar Bone Involvement** ▶ Relatively common.  
▶ The gingiva are normal.
- Other Sites of Involvement** ▶ Bones (skull, calvaria)  
▶ Skin  
▶ Gastrointestinal tract
- Main Clinical Features** ▶ The alveolar bone lesions are characterized by multiple osteomas that present as painless, hard masses covered by normal attached gingiva and alveolar mucosa (**Fig. 159**). Central osteomas in other sites of the jaws, multiple odontomas, multiple impacted supernumerary teeth and rarely benign oral soft tissue tumours may occur. It is important for dentists to diagnose the oral lesions of the syndrome as early as possible and to refer the patient to a gastroenterologist.  
▶ Multiple osteomas of the skull, paranasal sinuses and calvaria may also develop.  
▶ Gastrointestinal manifestations are common and appear as multiple polyps of the colon and rectum that are characterized by a high rate of malignant transformation into adenocarcinoma.  
▶ Skin lesions are epidermoid cysts, multiple fibromas and hyperpigmentation.
- Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.  
▶ Radiographic examination of the jaws, skull and intestinal tract.  
▶ Biopsy and histopathological examination of tumours and polyps.
- Differential Diagnosis** ▶ Isolated osteomas and exostoses (tori)  
▶ Odontogenic tumours  
▶ Peutz-Jeghers syndrome  
▶ Cowden's syndrome
- Treatment** ▶ Osteomas do not require excision unless they cause symptoms or cosmetic problems.  
▶ The intestinal polyps must be under observation by the specialist.

### Sturge-Weber Angiomatosis

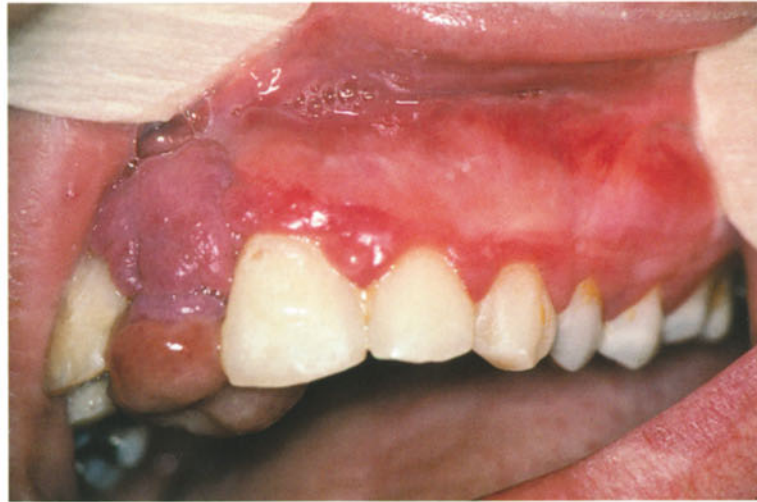
- Definition** ▶ Sturge-Weber angiomatosis or encephalotrigeminal angiomatosis is a rare sporadic congenital hamartomatous vascular disorder typically involving areas innervated by the trigeminal nerve.
- Etiology** ▶ Obscure; developmental abnormality
- Gingival Involvement** ▶ Relatively common
- Other Sites of Involvement** ▶ Oral mucosa  
▶ Skin  
▶ Eyes, central nervous system

**FIGURE 159**

Gardner's syndrome.  
Multiple osteomas of the  
alveolar bone

**FIGURE 160**

Sturge-Weber angiomatosis.  
Unilateral gingival haemangiomas

**FIGURE 161**

Sturge-Weber angiomatosis.  
Unilateral gingival haemangiomas





FIGURE 162

Sturge-Weber angiomas.  
Unilateral facial haemangiomas



- Main Clinical Features**
- ▶ Typically unilateral haemangiomas, usually capillary, on the attached gingiva and the alveolar mucosa may be seen occasionally leading to gingival hyperplasia and premature or rarely delayed eruption (Figs. 160, 161). The lesions have a bright red or purple colour and are usually flat. Dentists must be careful during tooth extraction and periodontal surgery so as to avoid bleeding complications.
  - ▶ Unilateral oral haemangiomas on the buccal mucosa, tongue, and lips may occur.
  - ▶ Unilateral facial haemangioma is the most characteristic and consistent feature, it is evident at birth and characteristically it is confined to the area supplied by the trigeminal nerve (Fig. 162)
  - ▶ Haemangiomas of the leptomeninges, brain calcification, epilepsy, ocular disorders may also be present.
- Diagnosis**
- ▶ The diagnosis is usually based on clinical criteria.
  - ▶ Histopathological examinations
  - ▶ CT scan.
- Differential Diagnosis**
- ▶ Solitary haemangioma
  - ▶ Klippel-Trenaunay-Weber syndrome
- Treatment**
- ▶ Laser therapy is recommended.

### Klippel-Trenaunay-Weber Syndrome

- Definition**
- ▶ Klippel-Trenaunay-Weber syndrome is an uncommon dysplastic vascular disorder.
- Etiology**
- ▶ Dysplastic malformation
- Gingival Involvement**
- ▶ Common
- Other Sites of Involvement**
- ▶ Oral mucosa
  - ▶ Skin
  - ▶ Eyes
  - ▶ Bone and soft tissues of extremities



**FIGURE 163**

Klippel-Trenaunay-Weber syndrome.  
Gingival haemangiomas leading  
to gingival overgrowth

**FIGURE 164**

Klippel-Trenaunay-Weber syndrome.  
Scleral pigmentation

**FIGURE 165**

Klippel-Trenaunay-Weber syndrome.  
Haemangiomas of the leg



- Main Clinical Features**
- ▶ The gingival lesions appear as capillary or cavernous haemangiomas occasionally leading to gingival overgrowth that may cover the crown of the teeth (**Fig. 163**). Premature tooth eruption and alveolar bone overgrowth may occur.
  - ▶ The oral mucosal lesions are also haemangiomas, particularly on the hard and soft palate.
  - ▶ Multiple facial haemangiomas, ocular disorders (scleral pigmentation, glaucoma, cataract, iris heterochromia), vascular masses that involve soft tissue and bone leading to asymmetrical enlargement of the extremities, haemangiomas in internal organs are common (**Figs. 164, 165**).
- Diagnosis**
- ▶ The diagnosis is based on clinical criteria.
- Differential Diagnosis**
- ▶ Isolated haemangiomas
  - ▶ Sturge-Weber syndrome
  - ▶ Maffucci's syndrome
- Treatment**
- ▶ Palliative.
  - ▶ Laser may improve skin lesions.

### Hereditary Epidermolysis Bullosa

- Definition**
- ▶ Hereditary epidermolysis bullosa is a rare chronic, recurrent, mechanobullous group of genetic mucocutaneous disorders.
- Etiology**
- ▶ Genetic. The exact etiology and pathogenesis are obscure.
- Classification**
- ▶ Three major forms and numerous subtypes are recognized:
    - The intraepidermal, type I (characterized by a non-scarring pattern and inherited as an autosomal and X-linked trait).
    - The junctional, type II (characterized by skin atrophy and inherited as an autosomal recessive trait).
    - The dermal, type III (characterized by skin atrophy and scarring and inherited as an autosomal dominant or recessive trait).
- Gingival Involvement**
- ▶ Common, particularly of types II and III
  - ▶ Non-dental plaque-induced gingival lesions, occasionally associated with attachment loss
- Other Sites of Involvement**
- ▶ Oral mucosa (tongue, buccal mucosa, palate, lips)
  - ▶ Other mucosae (eyes, larynx, oesophagus, genitals)
  - ▶ Skin, always (hands, feet, fingers, toes, knee, elbows, scale, nails), mainly in areas of mechanical friction
- Main Clinical Features**
- ▶ The gingival manifestations usually follow the skin lesions and may appear in the form of desquamative gingivitis or as localized or widespread bullae formation that rupture soon leaving painful erosions (**Figs. 166, 167**). Gingival recession and periodontal destruction may also occur in severe cases.
  - ▶ The oral mucosal lesions appear as bullae formation that soon rupture, leaving painful erosions or ulcerations that may occasionally lead to scarring. Microstomia, epithelial atrophy, elimination of tongue movement, elimination of vestibular and buccal sulci, lobulated hyperplastic nodules, pitted enamel and teeth caries may occur (**Fig. 168**).



**FIGURE 166**

Hereditary epidermolysis  
bullosa, type I.  
Solitary bulla formation  
on the alveolar mucosa

**FIGURE 167**

Hereditary epidermolysis  
bullosa, type II.  
Gingival erosions and enlargement

**FIGURE 168**

Hereditary epidermolysis  
bullosa, type III.  
Pitted enamel and tooth caries



**FIGURE 169**

Hereditary epidermolysis  
bullosa, type II.  
Bulla formation and scarring  
of the skin

**FIGURE 170**

Hereditary epidermolysis  
bullosa, type III.  
Scarring and deformities of the hand

**FIGURE 171**

Hereditary epidermolysis  
bullosa, type II.  
Nail dystrophy





- ▶ The skin manifestations are characterized by bulla formation, ulcerations, scarring, deformities, milia, nail dystrophy and several others (Figs. 169–171).
- ▶ The clinical manifestations of the disorder present early in life (infants and children) and the severity of the skin and mucosal lesions depend on the form of the disease.

- Diagnosis**
- ▶ The diagnosis is based on clinical and laboratory criteria.
  - ▶ Biopsy and histopathological examination.
  - ▶ Immunofluorescence test.
  - ▶ Electron microscopic examination.

- Differential Diagnosis**
- ▶ Cicatricial pemphigoid
  - ▶ Bullous pemphigoid
  - ▶ Pemphigus vulgaris
  - ▶ Linear IgA disease

- Treatment**
- ▶ Avoid pressure on the gingiva and oral mucosa.
  - ▶ Good oral hygiene.
  - ▶ Topical corticosteroid application.
  - ▶ Systemic corticosteroid in severe lesions.

### Chondroectodermal Dysplasia

- Definition**
- ▶ Chondroectodermal dysplasia or Ellis-van Creveld syndrome is an uncommon disorder of the osteochondrodysplasia group of defects.

- Etiology**
- ▶ Genetic. It is inherited as an autosomal recessive trait.

- Gingival Involvement**
- ▶ Common

- Other Sites of Involvement**
- ▶ Teeth, skeleton, eyes, heart, central nervous system
  - ▶ Hair and nails

- Main Clinical Features**
- ▶ Fusion of the middle portion of the lower or upper lip to the gingiva, leading to the disappearance of the mucolabial fold and multiple frenular is the most characteristic oral manifestation (Figs. 172, 173). Oligodontia, and small teeth with conical crowns may occur.
  - ▶ Chondrodysplasia of the long bones, bilateral polydactyly, shortened extremities, thin and sparse hair, dystrophic nails, heart defects and genital abnormalities are common (Fig. 174).

- Diagnosis**
- ▶ The diagnosis is mainly based on clinical criteria.
  - ▶ Radiographic examination.

- Differential Diagnosis**
- ▶ Orofaciodigital syndrome
  - ▶ Other chondrodysplasia syndromes
  - ▶ Hereditary epidermolysis bullosa, type III

- Treatment**
- ▶ Supportive.

FIGURE 172

Chondroectodermal dysplasia.  
Multiple frenulum and disappearance  
of the mucolabial fold



FIGURE 173

Chondroectodermal dysplasia.  
Multiple gingivolabial frenulum



FIGURE 174

Chondroectodermal dysplasia.  
Polydactyly and nail dystrophy



### Focal Dermal Hypoplasia Syndrome

- Definition** ▶ Focal dermal hypoplasia syndrome or Goltz-Gorlin syndrome is an uncommon disorder characterized by multiple abnormalities.
- Etiology** ▶ Genetic. It is inherited as an X-linked dominant trait. However, most of the reported patients are sporadic cases.
- Gingival Involvement** ▶ Rare
- Other Sites of Involvement** ▶ Oral mucosa (tongue, palate, buccal mucosa, lips) and teeth  
▶ Skin, hair and nails  
▶ Musculoskeletal and central nervous systems, eyes
- Main Clinical Features** ▶ The gingival lesions are papillomas (**Fig. 175**).  
▶ Multiple papillomas on the oral mucosa are present in about 50 % of the cases. Oligodontia, hypodontia, enamel dysplasia and malocclusion, oral clefts are common.  
▶ Skin manifestations include atrophic linear lesions, atrophic and hyperpigmented plaques, telangiectasias and soft yellowish nodules (**Fig. 176**). Sparse hair and dystrophic nails may be observed.  
▶ Syndactyly, brachydactyly, oligodactyly or polydactyly, clinodactyly, extremity asymmetry, scoliosis, and microcephaly may occur.  
▶ Eye anomalies, learning disability are less common findings.
- Diagnosis** ▶ The diagnosis is mainly based on clinical criteria.  
▶ Biopsy and histopathological examination of skin and oral lesions.
- Differential Diagnosis** ▶ Multiple oral papillomas  
▶ Multiple oral condylomas acuminata  
▶ Focal epithelial hyperplasia  
▶ Incontinentia pigmenti
- Treatment** ▶ Surgical excision of oral papillomas  
▶ Supportive for the skin and other lesions

**FIGURE 175**

Focal dermal hypoplasia syndrome.  
Multiple papillomas on the gingiva  
and the palate





FIGURE 176

Focal dermal hypoplasia syndrome.  
Skin atrophy



### Orofaciodigital Syndrome Type I

**Definition** ▶ Orofaciodigital syndrome, type I is an uncommon disorder in the group of orofacioidigital syndromes.

**Etiology** ▶ Genetic. It is inherited as an X-linked dominant trait, limited to females and fatal in males.

**Gingival Involvement** ▶ Common

**Other Sites of Involvement** ▶ Oral mucosa, teeth  
▶ Facies, digits  
▶ Skin

**Main Clinical Features** ▶ The gingival involvement characterized by multiple hyperplastic fibrous bands traversing the gingivolabial and gingivobuccal fold, hypertrophy and shortening of the frenulum of the lips and tongue (**Figs. 177, 178**). These may result in hyperplastic gingivitis and gingival recession.  
▶ The oral mucosal lesions include multilobed or bifid tongue, with multiple hamartomas, lips and palate clefts, supernumerary teeth, teeth malposition and missing teeth, hypoplastic mandible.  
▶ Frontal bossing, ocular hypertelorism, short upper lip, broad nasal root, asymmetry of the nostrils, polydactyly, brachydactyly, clinodactyly and syndactyly are common.  
▶ The skin manifestations include xeroderma, milia, alopecia and sparse hair.  
▶ Learning disability and polycystic kidneys may occasionally occur.

**Diagnosis** ▶ The diagnosis is based on clinical criteria.

**Differential Diagnosis** ▶ Orofaciodigital syndrome type II  
▶ Chondroectodermal dysplasia  
▶ Oculodentodigital syndrome

**Treatment** ▶ Surgical reconstruction of oral frenulum and gingivolabial and gingivobuccal folds  
▶ Symptomatic for the rest of lesions

**FIGURE 177**

Orofaciodigital syndrome, type I.  
Multiple gingivolabial frenulum

**FIGURE 178**

Orofaciodigital syndrome, type I.  
Gingivitis and gingival hyperplasia



### Ehlers-Danlos Syndrome

**Definition** ▶ Ehlers-Danlos syndrome includes a group of connective tissue disorders that are characterized by defects in collagen synthesis.

**Etiology** ▶ Genetic. It is mainly inherited as an autosomal dominant trait.

**Classification** ▶ On the basis of clinical features and the mode of inheritance, ten types of the disorders are recognized.

**Periodontal Involvement** ▶ Relatively common. Mainly types III, IV and VIII have an increased susceptibility to periodontitis.  
▶ Possibly multigene disorder influenced by dental plaque level.  
▶ Periodontitis associated with genetic disorder.

FIGURE 179

Ehlers-Danlos syndrome.  
Generalized periodontitis



**Other Sites of Involvement**

- ▶ Oral mucosa
- ▶ Skin
- ▶ Joints
- ▶ Heart

**Main Clinical Features**

- ▶ Periodontal lesions usually appear in the form of severe generalized aggressive periodontitis with clinical attachment loss and bone destruction, leading to premature loss of permanent teeth (**Fig. 179**). Gingival bleeding is also common.
- ▶ The oral mucosa manifestations include thin and fragile mucosa, prolonged bleeding on minor trauma and decreased wound healing. About 50% of patients have the ability to touch their nose with the tip of the tongue, compared with 10% of normal people. Temporomandibular joint hypermobility, multiple pulp stones, enamel, dentin and cementum defects may occur.
- ▶ The skin manifestations include thin and fragile skin, hypermobility, haematomas, scarring formation, especially over the knees, elbows and chin.
- ▶ Joint hypermobility, cardiovascular and ocular abnormalities may occur, especially mitral valve prolapse.

**Diagnosis** ▶ The diagnosis is based on clinical criteria.

**Differential Diagnosis**

- ▶ Aggressive periodontitis associated with other genetic disorders
- ▶ Marfan's syndrome
- ▶ Cutis laxa

**Treatment**

- ▶ Dental plaque control
- ▶ Symptomatic



### Hereditary Haemorrhagic Telangiectasia

- Definition** ▶ Hereditary haemorrhagic telangiectasia, or Osler-Rendu-Weber disease, is a relatively rare disorder characterized by permanent small dilations of vessels.
- Etiology** ▶ Genetic. It is inherited as an autosomal dominant trait.
- Gingival Involvement** ▶ Rare
- Other Sites of Involvement** ▶ Oral mucosa (tongue, lips, buccal mucosa, palate)  
▶ Other mucosae (nasal, oesophagus, genital)  
▶ Skin  
▶ Internal organ (liver, spleen, gastrointestinal tract, lungs)
- Main Clinical Features** ▶ The gingival lesions appear as small (usually 1–3 mm in size), localized or multiple, bright red macules or nodules that characteristically disappear when pressure is applied (Figs. 180, 181). Gingival recurrent bleeding, usually during or after tooth brushing, is frequent.  
▶ The oral mucosal manifestations are characterized by multiple telangiectasias and rarely bleeding (Fig. 182).  
▶ Persistent recurrent nose bleeding is usually the first and common clinical sign of the disease.  
▶ Multiple telangiectasias on the skin and gastrointestinal tract are common.  
▶ Liver and spleen vascular abnormalities may be less frequent.  
▶ The disease usually manifests during the second or third decade of life.
- Diagnosis** ▶ The diagnosis is mainly based on clinical criteria.  
▶ Biopsy and histopathological examination.
- Differential Diagnosis** ▶ CREST syndrome  
▶ Fabry's disease  
▶ Maffucci's syndrome
- Treatment** ▶ Usually no treatment is required for oral lesions.  
▶ If the lesions often bleed, surgical excision or laser therapy is suggested.

FIGURE 180

Hereditary haemorrhagic telangiectasia.  
Solitary red lesion on the gingiva

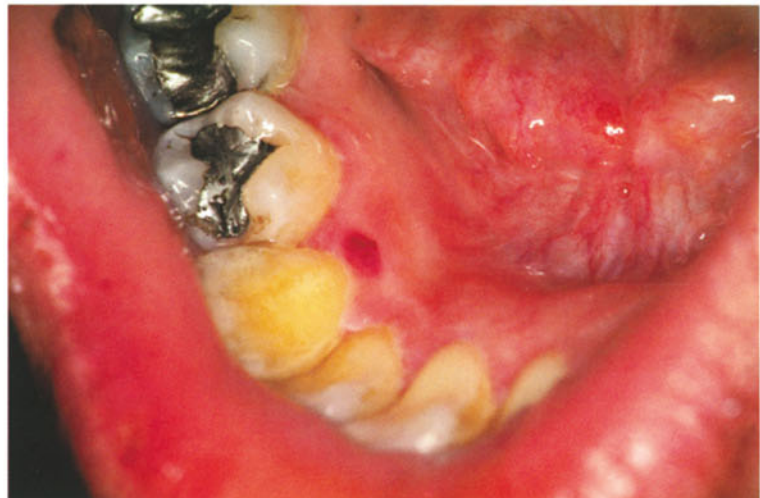


FIGURE 181

Hereditary haemorrhagic  
telangiectasia.  
Solitary red lesion on the  
alveolar mucosa

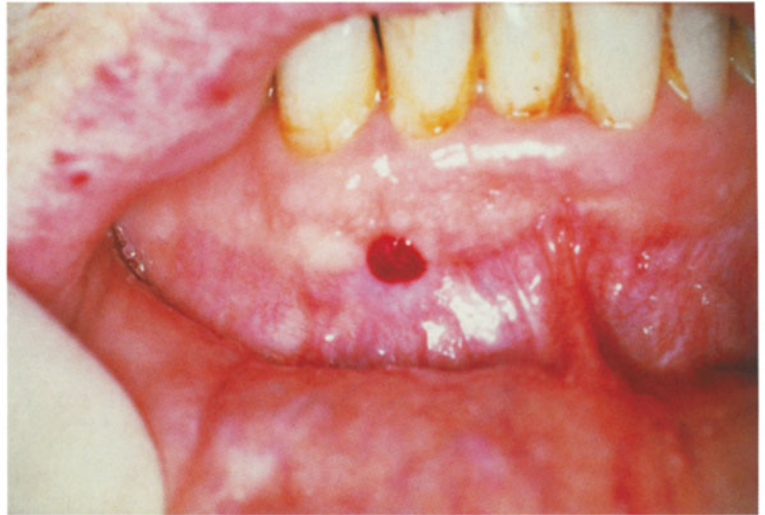


FIGURE 182

Hereditary haemorrhagic  
telangiectasia.  
Multiple red lesions on the tongue



## Cystic Fibrosis

- Definition** ▶ Cystic fibrosis is a relatively common, multisystemic, life-threatening disorder characterized by a dysfunction of the exocrine glands.
- Etiology** ▶ Genetic. It is inherited as an autosomal recessive trait. The pathological gene known as the *CFTR* gene, is located on the long arm of chromosome 7, and causes a disturbance in chloride ion transport across the cell membrane.
- Gingival Involvement** ▶ Very rare  
▶ Dental plaque-induced gingivitis associated with xerostomia
- Other Sites of Involvement** ▶ Oral mucosa (lips), relatively common  
▶ Pulmonary and gastrointestinal tracts  
▶ Liver, pancreas  
▶ Sweat glands
- Main Clinical Features** ▶ The gingival lesions are uncommon and present as a non-specific diffuse erythema on both free and attached gingiva (**Fig. 183**). The gingival lesions may be due to oral dryness and the dental plaque.  
▶ The oral manifestations include lip enlargement, erythema of the oral mucosa, and occasionally mild xerostomia.  
▶ Chronic coughing, pulmonary infections, shortness of breath, bronchitis, haemoptysis are common.  
▶ Malabsorption, bulky, greasy stools, and failure to gain weight even with adequate food intake are common.  
▶ Liver and pancreatic insufficiency, sweating with salty taste are also common.  
▶ The disease starts early in infancy.
- Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.  
▶ Biopsy and histopathological examination of the lip.  
▶ Elevated chloride, sodium and potassium levels in sweat.  
▶ Absence of pancreatic enzymes in the intestinal fluid.

**FIGURE 183**

Cystic fibrosis.  
Gingivitis





- Differential Diagnosis**
- ▶ Gingivitis associated with dental plaque only
  - ▶ Lipoid proteinosis
  - ▶ Cheilitis glandularis and granulomatosa

- Treatment**
- ▶ Dental plaque control for the gingival lesions.
  - ▶ Palliative high-calorie diet indicated for the systemic manifestations.

### Down's Syndrome

- Definition**
- ▶ Down's syndrome, or trisomy 21, is a common disorder with variable clinical manifestations. The incidence in the general population is 1 in 600–800 live births.

- Etiology**
- ▶ Chromosomal disorder of chromosome 21.
  - ▶ Most patients with Down's syndrome exhibit full trisomy 21. Few cases are mosaic and fewer are the result of translocation.

- Periodontal Involvement**
- ▶ Common
  - ▶ Dental plaque-induced gingival lesions and frequently periodontitis associated with genetic disorders

- Other Sites of Involvement**
- ▶ Oral cavity, common
  - ▶ Craniofacies, thorax, abdomen, pelvis, hands and feet

- Main Clinical Features**
- ▶ Clinically the gingiva are red, swollen and bleed easily. (**Fig. 184**). Gingival hyperplasia may occur due to chronic mouth breathing, poor oral hygiene and other local irritating factors. The gingivitis frequently progresses to generalized early periodontitis, which commences in the deciduous dentition and continues into the permanent dentition. Bone loss, mobility and migration of the teeth are common. All these predispose to the premature loss of teeth.
  - ▶ Macroglossia, fissured and geographic tongue, high-arched palate, cleft palate, delayed tooth eruption and hypoplastic teeth are the most frequent oral lesions.
  - ▶ Follicular hyperkeratosis, facial flushing, alopecia areata, and dry skin are common.
  - ▶ Polydactyly, syndactyly, clinodactyly, hyperextensibility of joints, other skeletal abnormalities, congenital heart anomalies, hormonal disturbances, learning disability, mongoloid slanting of the eyes, short ears, and flat face are common findings.

- Diagnosis**
- ▶ The diagnosis is usually based on clinical criteria.
  - ▶ Chromosomal analysis.

- Differential Diagnosis**
- ▶ Cyclic neutropenia
  - ▶ Agranulocytosis
  - ▶ Leukaemias
  - ▶ Glycogen storage disease type 1b

- Treatment**
- ▶ Good oral hygiene and plaque control reduce the gingival and periodontal lesions.

FIGURE 184

Down's syndrome.  
Severe gingivitis



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### Tuberculosis

- Definition** ▶ Tuberculosis is a chronic granulomatous infectious disease.
- Etiology** ▶ *Mycobacterium tuberculosis*  
▶ Non-dental plaque-induced gingival lesions
- Gingival Involvement** ▶ Rare
- Other Involvement** ▶ Oral mucosa (tongue, palate, uvula, buccal, lips), rare  
▶ Lungs, almost always
- Main Clinical Features** ▶ The gingival lesions may present as chronic, irregular, usually painless, ulcers with vegetating red surface or covered by a grey-yellowish exudate (Figs. 185, 186). Vegetating diffuse gingival enlargement may also occur.  
▶ The oral mucosal lesions manifest as an irregular, vegetating ulcer with a thin border. The surrounding tissue is slightly indurated with inflammation. Regional lymphadenopathy accompanies the oral lesions.  
▶ Tuberculous osteomyelitis of the jaws may rarely occur.  
▶ Gingival and oral manifestations usually represent secondary infection from the initial pulmonary infection and the oral prevalence varies from 0.5% to 1.5%.  
▶ Rarely, oral tuberculosis may be the only manifestation of otherwise silent tuberculosis.  
▶ Low-grade fever, anorexia, malaise, cough, night sweats are early and common symptoms. Chest pain and haemoptysis are also common.
- Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.  
▶ Biopsy and histopathological examination of oral lesions.  
▶ Chest radiographs.  
▶ Mycobacterial examination or culture of infected sputum and tissues.  
▶ Mantoux tuberculin skin test.
- Differential Diagnosis** ▶ Traumatic ulcer  
▶ Squamous cell carcinoma  
▶ Primary and secondary syphilis  
▶ Major aphthous ulcer  
▶ Non-Hodgkin's lymphoma  
▶ Wegener's granulomatosis  
▶ Pyostomatitis vegetans
- Treatment** ▶ Antituberculous multiple drug regimens that must be left to the specialist

FIGURE 185

Tuberculosis.  
Small ulcer on the upper gingiva



FIGURE 186

Tuberculosis.  
Vegetating, irregular ulcer  
on the lower gingiva





## Syphilis

- Definition** ▶ Syphilis is a sexually transmitted disease involving several organs and systems.
- Etiology** ▶ *Treponema pallidum*
- Classification** ▶ Congenital.  
▶ Acquired. Acquired syphilis develops in three stages:
  - Primary.
  - Secondary.
  - Tertiary.
- Gingival Involvement** ▶ Rare  
▶ Non-dental plaque-induced gingival lesions
- Other Involvement** ▶ Oral mucosa, common. Approximately 5%–10% of the primary and over of 40% of secondary syphilis appears on the oral cavity.  
▶ Genitalia, very common.  
▶ Skin, common (secondary and tertiary stages).
- Main Clinical Features** ▶ Primary syphilis is characterized by the development of chancre at the site of inoculation of *treponema pallidum*. Chancre begins as an inflammatory papule that ulcerates to form a painless round ulcer with a raised border and indurated base. Chancre on the gingiva is extremely rare (Figs. 187, 188). The lips, tongue, soft palate, and tonsils are the most common oral sites affected. Regional lymph node enlargement is a constant finding. Direct orogenital contact is necessary for the development of an oral chancre.  
▶ Secondary syphilis begins 6–8 weeks after the appearance of the chancre. Oral lesions are common in this stage and are mainly characterized by the appearance of mucous patches, which are painless, slightly raised, oval or round erosions or superficial ulcers covered by a greyish-white membrane. Gingival involvement is relatively rare (Fig. 189). The tongue, buccal mucosa, palate, tonsils and lips are most frequently affected. The classic signs and symptoms of secondary syphilis are headache, malaise, fever, generalized painless lymph node enlargement and maculopapular skin eruption.  
▶ Tertiary syphilis (late syphilis) develops in about 30% of untreated patients, and is characterized by multisystem involvement and broad spectrum of manifestations. The gumma is the most characteristic oral lesion of this stage, and the hard palate is the most common site of involvement, followed by tongue and lips. Gingiva is not involved.
- Diagnosis** ▶ The clinical diagnosis of syphilis should be confirmed by laboratory tests.  
▶ Serological tests.  
▶ Darkfield microscopy.
- Differential Diagnosis** ▶ Herpetic oral lesions  
▶ Traumatic oral lesions  
▶ Aphthous ulcer  
▶ Candidiasis  
▶ Erythema multiforme  
▶ Infectious mononucleosis
- Treatment** ▶ Penicillin or erythromycin, but it must be left to the physician

FIGURE 187

Primary syphilis.  
Small chancre on the border of  
attached gingiva and alveolar mucosa



FIGURE 188

Primary syphilis.  
Large chancre on the edentulous  
alveolar mucosa



FIGURE 189

Secondary syphilis.  
Mucous patches on the lower gingiva





## Actinomycosis

- Definition** ▶ Actinomycosis is a chronic granulomatous infectious disease.
- Etiology** ▶ Mainly *Actinomyces israelii* and rarely other species
- Gingival Involvement** ▶ Rare  
▶ Non-dental plaque-induced gingival lesions
- Other Involvement** ▶ Jaws  
▶ Oral mucosa  
▶ Cervical lymph nodes
- Main Clinical Features** ▶ Oral manifestations are part of the cervicofacial actinomycosis.  
▶ Gingival lesions are extremely rare and may present as localized atypical gingivitis or more commonly as painless alveolar bone swelling covered by slightly erythematous gingiva, usually with draining sinuses (**Fig. 190**).  
▶ Involvement of the jaws is common but soft tissues (tongue, palate, buccal) may be less frequently involved. Painless, slow-growing, hard swelling is the classic feature of the disease, usually followed by multiple abscesses that drain to the surface by sinus tracts (**Fig. 191**). The exudation from the sinus tracts typically contains the characteristic sulphur granules that are colonies of organisms.
- Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.  
▶ Direct bacteriological examination and culture.  
▶ Indirect immunofluorescence.  
▶ Biopsy and histopathological examination.
- Differential Diagnosis** ▶ Periodontal and dental abscess  
▶ Tuberculosis  
▶ Systemic mycoses
- Treatment** ▶ Penicillin, erythromycin or tetracyclines.  
▶ Surgery is necessary in most cases.

**FIGURE 190**

Actinomycosis.  
Draining sinus on the gingiva





FIGURE 191

Actinomycosis.  
Multiple sinus tracts on the skin



### Gonococcal Infection

**Definition** ▶ Gonococcal infection is a sexually transmitted disease that primarily involves the urinary tract and less often the respiratory tract.

**Etiology** ▶ *Neisseria gonorrhoeae*

**Gingival Involvement** ▶ Rare  
▶ Non-dental plaque-induced gingival lesions

**Other Involvement** ▶ Oral mucosa, relatively rare. It is more common among homosexuals and heterosexuals practising oral sex.  
▶ Urethra, anus, cervix.

**Main Clinical Features** ▶ Gingival involvement may present as non-specific erythema and slight oedema with occasionally painful erosions (**Fig. 192**).  
▶ The oral mucosal manifestations present as fiery erythema with or without superficial ulceration, usually covered with a greyish or yellowish exudate. The tonsil and soft palate are the most commonly affected sites followed by the tongue and buccal mucosa.

**Diagnosis** ▶ The clinical diagnosis of oral gonococcal infection should always be confirmed by laboratory tests.  
▶ Culture and microscopic identification of microorganism.  
▶ Fluorescent antibody test.

**Differential Diagnosis** ▶ Dental plaque-induced gingivitis  
▶ Desquamative gingivitis  
▶ Herpetic gingivitis and stomatitis  
▶ Granulomatous gingivitis  
▶ Streptococcal gingivitis and stomatitis

**Treatment** ▶ Frequently oral lesions are self-limited.  
▶ Systemic penicillin or tetracycline.

FIGURE 192

Gonococcal gingivitis  
presenting as localized erythema  
and slight oedema



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### Measles

- Definition** ▶ Measles is a highly communicable infectious disease of childhood.
- Etiology** ▶ RNA paramyxovirus belonging to the *Morbillivirus* genus
- Gingival Involvement** ▶ Rare
- Other Involvement** ▶ Oral mucosa, relatively rare  
▶ Skin, always
- Main Clinical Features** ▶ The gingival lesions may present as usually isolated, small round ulcers on the attached gingiva or the alveolar mucosa and occasionally as gingival erythema (**Figs. 193, 194**). The gingival lesions are not diagnostic.  
▶ The oral mucosal lesions present as Koplik's spots (on the buccal mucosa), redness, petechiae and small round ulcers.  
▶ The skin manifestations present as maculopapular dull red eruptions.  
▶ Constitutional symptoms such as fever, chills, cough, photophobia, conjunctivitis and myalgias begin 3–4 days prior to skin eruption.
- Diagnosis** ▶ The diagnosis is based on clinical criteria.
- Differential Diagnosis** ▶ Varicella  
▶ Herpetic gingivostomatitis  
▶ Herpetiform ulcers  
▶ Infectious mononucleosis
- Treatment** ▶ No treatment is needed for the gingival and oral lesions.  
▶ The general management of patients must be left to the paediatrician.



**FIGURE 193**

Measles.  
Small round ulcer on the  
attached gingiva

**FIGURE 194**

Measles.  
Gingival erythema



### Chickenpox

**Definition** ▶ Chickenpox or varicella is a highly contagious, self-limited primary infection, mainly in children.

**Etiology** ▶ Varicella-zoster virus or HHV-3

**Gingival Involvement** ▶ Rare

**Other Involvement** ▶ Oral mucosa (palate, buccal, lip), relatively rare  
▶ Skin (face, head, neck), always

**Main Clinical Features** ▶ The gingival lesion present usually as diffuse erythema and isolated small vesicles that rupture quickly leaving round shallow ulcerations on an erythematous area (**Fig. 195**).  
▶ The oral mucosal lesions also present as small vesicles, which rupture quickly leaving small ulcerations.

**FIGURE 195**

Chickenpox.  
Diffuse gingival erythema



- ▶ The skin manifestations include a pruritic red maculopapular rash, vesicles and pustules that rupture, crust and later exfoliate.
- ▶ The prodromal symptoms are malaise, fever, anorexia and irritability.

#### Diagnosis

- ▶ The diagnosis is mainly based on clinical criteria and rarely on laboratory tests.
- ▶ Tzanck smear examination.
- ▶ Polymerase chain reaction (PCR) detection of varicella-zoster virus DNA may be useful.

#### Differential Diagnosis

- ▶ Herpes simplex oral infection
- ▶ Minor aphthous ulcers
- ▶ Measles

#### Treatment

- ▶ No treatment is needed for oral lesions.
- ▶ Supportive treatment should be left to the paediatrician.

### Herpes Zoster

#### Definition

- ▶ Herpes zoster is an acute viral disease caused by reactivation of a latent varicella-zoster virus that is transported via sensory nerve axons to the mucosae and/or the skin.

#### Etiology

- ▶ Reactivation of varicella-zoster virus, particular in immunocompromised patients

#### Gingival Involvement

- ▶ Relatively common. Gingival and oral mucosal lesions may occur when second or third divisions of the trigeminal nerve are involved.

#### Other Involvement

- ▶ Oral mucosa (palate, buccal, tongue, lips)
- ▶ Skin (face)

#### Main Clinical Features

- ▶ The gingival lesions present as unilateral clusters of vesicles which in 2–3 days rupture leaving painful ulcerations with irregular borders (Figs. 196, 197). Necrosis of the periodontium and alveolar bone may be seen. The ulcers usually heal without scarring in 2–3 weeks.

FIGURE 196

Herpes zoster.  
Ulceration of the lower attached  
gingiva in mandibular zoster



FIGURE 197

Herpes zoster.  
Diffuse erythema and clusters of  
vesicles on the gingiva and the  
palatal mucosa in maxillary zoster



FIGURE 198

Herpes zoster.  
Typical unilateral lesions on the face  
in maxillary zoster





- ▶ Similar unilateral lesions may appear on other oral sites, depending on trigeminal division involvement.
- ▶ An itching sensation and pain, which may simulate pulpitis, may precede oral lesions.
- ▶ The skin manifestations (trigeminal involvement) appear on the face and are characterized by unilateral clusters of maculopapules on an erythematous base, which soon form vesicles and in 2–3 days evolve into pustules. The pustules form within 5–8 days' crust and persist for 10–20 days (**Fig. 198**). New lesions continue to appear for several days. Tenderness and pain is always present.
- ▶ Constitutional symptoms such as fever, malaise and headache are common.
- ▶ Post-herpetic trigeminal neuralgia is the most common complication.

- Diagnosis**
- ▶ The diagnosis is based on clinical criteria and laboratory tests may be necessary only in atypical cases:
    - Viral culture.
    - Cytology smear.
    - Serological tests.
    - Viral DNA studies.

- Differential Diagnosis**
- ▶ Oral herpes simplex infection
  - ▶ Necrotizing ulcerative gingivitis and periodontitis
  - ▶ Erythema multiforme
  - ▶ Leukaemia
  - ▶ Agranulocytosis

- Treatment**
- ▶ Antiviral drugs (valacyclovir, famciclovir, aciclovir)
  - ▶ Analgesics and sedatives

### Infectious Mononucleosis

- Definition**
- ▶ Infectious mononucleosis is an acute self-limited infectious disease that is transmitted by saliva or blood.

- Etiology**
- ▶ Epstein-Barr virus (or HHV-4)

- Gingival Involvement**
- ▶ Rare

- Other Involvement**
- ▶ Oral mucosa (soft palate, uvula, buccal), relatively common
  - ▶ Skin (rash)
  - ▶ Lymph nodes

- Main Clinical Features**
- ▶ The gingival lesions may present as localized or diffuse erythema and less frequently as small ulcerations, particularly at the interdental papillae, mimicking necrotizing ulcerative gingivitis (**Figs. 199, 200**).
  - ▶ The oral mucosal manifestations include palatal petechiae, faucial oedema, tonsillar exudate, pericoronitis and occasionally ulcerations.
  - ▶ The skin lesions present as a maculopapular eruption.
  - ▶ Generalized lymphadenopathy, profound malaise, sore throat, fever, anorexia and headache are common signs and symptoms.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests:
    - Monospot test.
    - Paul-Bunnell test.

FIGURE 199

Infectious mononucleosis.  
Diffuse erythema on the  
upper gingiva and lip



FIGURE 200

Infectious mononucleosis.  
Localized erythema on the  
lower gingiva and necrotizing  
gingivitis



- Differential Diagnosis**
- ▶ Herpetic gingivostomatitis
  - ▶ Streptococcal gingivitis
  - ▶ Necrotizing ulcerative gingivitis
  - ▶ Leukaemia
  - ▶ Aplastic anaemia

- Treatment**
- ▶ Supportive.

### Hand-Foot-and-Mouth Disease

- Definition** ▶ Hand-foot-and-mouth disease is an acute virus infection that usually affects children and young adults.
- Etiology** ▶ Coxsackie virus A16 and occasionally other types (A<sub>4</sub>, A<sub>5</sub>, A<sub>9</sub>, A<sub>10</sub>) are responsible.
- Gingival Involvement** ▶ Rare
- Other Involvement** ▶ Oral mucosa (buccal, labial, tongue, soft palate, uvula), common  
▶ Skin (hand, foot, back, buttocks), common
- Main Clinical Features** ▶ The gingival lesions are characterized by small vesicles that soon rupture, leaving slightly painful, shallow ulcers surrounded by a red halo (**Fig. 201**).  
▶ Similar lesions appear on the oral mucosa (**Fig. 202**).  
▶ The oral lesions are usually multiple (3–20), 2–6 mm in diameter.  
▶ The skin lesions present as multiple small vesicles (**Fig. 203**).  
▶ Low-grade fever, malaise, headache precedes the oral and skin lesions.  
▶ The disease lasts 5–8 days.
- Diagnosis** ▶ The diagnosis is based on clinical criteria.  
▶ Viral cultures and serological investigation needed only in doubtful cases.
- Differential Diagnosis** ▶ Aphthous ulcers  
▶ Herpetic stomatitis  
▶ Herpangina  
▶ Erythema multiforme
- Treatment** ▶ Symptomatic

**FIGURE 201**

Hand-foot-and-mouth disease.  
Small shallow ulcers on the gingiva





FIGURE 202

Hand-foot-and-mouth-disease.  
Multiple ulcers on the tongue



FIGURE 203

Hand-foot-and mouth-disease.  
Multiple small vesicles on the skin



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The spectrum of oral clinical signs and symptoms in HIV-infected patients is wide and its study is of great importance. Information concerning the nature, clinical and laboratory significance, and treatment of the oral lesions in HIV-infected patients has accumulated during the last 2 decades. Indeed, oral lesions in this group of patients have been used as indicators and early clinical signs not only of HIV disease but also of the disease progression and as criteria for entry into clinical trials.

It should be stressed that the oral lesions in HIV infection are indicative of and not truly specific for HIV disease, as several of them can also be found in non-HIV-infected patients. They should always be regarded as markers of HIV disease in correlation with the medical history and with other clinical and laboratory studies, particularly in cases where HIV status is unknown.

The oral lesions are mainly a result of cellular immunodeficiency due to HIV infection and are etiologically divided into five major groups. These include:

1. Infections (bacterial, viral, fungal)
2. Neoplasms
3. Drug reactions
4. Neurological manifestations
5. Lesions of unknown cause

These lesions may represent early or late manifestations of HIV disease, they may be common or rare, and the oral clinician should be familiar with all of them. The oral lesions are classified according to their relationship to HIV-infection into three groups:

- ▶ Group I, lesions that are strongly associated with HIV infection.
- ▶ Group II, lesions less commonly associated with HIV infection.
- ▶ Group III lesions are sometimes seen in HIV infection.

It should be emphasized that a dramatic reduction of oral lesions, including periodontal manifestations, has been observed the last 5 years in HIV-infected patients due to successful therapeutic regimens with the new anti-HIV drugs, resulting in improvement in the immunological status of patients.

It is of great importance for the periodontist to be familiar with periodontal manifestations of HIV infection, as the periodontium is a crucial anatomical region in dental practice.

The most common gingival lesions follow.



FIGURE 204

HIV infection.  
Necrotizing ulcerative gingivitis



### Bacterial Infections

#### Necrotizing Ulcerative Gingivitis (see also p. 93)

- ▶ Necrotizing ulcerative gingivitis (NUG) is a common early manifestation in HIV-infected patients, with a prevalence rate of 4%–16%. A high possibility for HIV infection exists, particularly if high-risk behaviour is associated with necrotizing ulcerative gingivitis.

#### Main Clinical Features

- ▶ Clinically, the onset of the disease is either acute or subacute and it presents with fiery red and swollen gingiva and a characteristic destruction of one or more interdental papillae. In the acute stage, ulceration, necrosis and sloughing of both the margin of the gingiva and the tip of the interdental papillae are common findings (**Fig. 204**).
- ▶ Pain, bleeding and halitosis are also common symptoms. The anterior gingiva are most frequently affected.
- ▶ Necrotizing ulcerative gingivitis should be limited to lesions only involving gingival tissue with no loss of periodontal attachment.

#### Diagnosis

- ▶ Diagnosis is made on clinical criteria. Necrotizing ulcerative gingivitis may rarely progress to necrotizing stomatitis in HIV-infected patients. The latter is characterized by localized or generalized acute, painful, ulcerative and necrotic lesions of the oral mucosa spread beyond the gingiva.

#### Treatment

- ▶ In the acute phase, treatment of NUG should include metronidazole, for 6–8 days and topical mouthwashes releasing oxygen.
- ▶ Conventional periodontal treatment (plaque-control measures, root planing, scaling) should follow.

#### Necrotizing Ulcerative Periodontitis

- ▶ Necrotizing ulcerative periodontitis (NUP) is the most severe form of periodontal disease associated with HIV infection and it develops in patients with marked immunosuppression. The prevalence of NUP is

FIGURE 205

HIV infection.  
Necrotizing ulcerative periodontitis



FIGURE 206

HIV infection.  
Localized necrotizing ulcerative  
periodontitis



FIGURE 207

HIV infection.  
Severe periodontitis presenting with  
severe oedema, multiple abscesses  
and bleeding





5 % or less. This low prevalence may be related to the improved recent medical management of HIV infection.

- Main Clinical Features**
- ▶ NUP is characterized by gingival ulceration and necrosis and a rapid progressive destruction of the periodontal attachment and bone with loosening of the teeth (Figs. 205, 206).
  - ▶ Spontaneous bleeding, deep-seated pain, halitosis, erythema and oedema are prominent features (Fig. 207).
  - ▶ The lesions may be localized or less often generalized and are usually not associated with deep pocket formation, because extensive gingival necrosis often coincides with loss of crestal alveolar bone.
- Treatment**
- ▶ Treatment of NUP includes systemic administration of metronidazole or tetracycline in association with topical antibacterial mouthwashes.
  - ▶ Conventional periodontal treatment maintenance and prevention should follow. Relapses are common.

### Bacillary Angiomatosis

- ▶ Bacillary angiomatosis is a recently described bacterial infection due to *Bartonella henselae* and *B. quintana*. It is characterized by vascular proliferation and occurs more frequently in HIV-infected patients and rarely in other immunosuppressed individuals or patients with malignancies. Skin, visceral and rarely gingival and oral lesions may occur. Oral bacillary angiomatosis should be regarded as an important prediction marker for HIV infection.
- Main Clinical Features**
- ▶ The oral lesions present as an asymptomatic, erythematous and slightly elevated soft tissue, plaque, or nodule (Fig. 208). The gingiva, palate and the tongue are the most commonly affected sites.
  - ▶ Clinical diagnosis is difficult because the lesion resembles Kaposi's sarcoma, pyogenic granuloma, pregnancy granuloma, peripheral giant cell granuloma, haemangioma, leiomyoma, brown giant cell tumour, amyloidosis and other vascular lesions.
  - ▶ The definitive diagnosis is based on histological and EM criteria.
- Treatment**
- ▶ Treatment consists of systemic erythromycin administration.
  - ▶ Tetracycline or doxycycline may also be used.

### Other Bacterial Infections

- ▶ Oral syphilis is a relatively common infection occurring in HIV-infected patients. Both primary and secondary syphilis can occur in the oral cavity and rarely, on the gingiva. In patients with HIV disease, the lesions may have a malignant and protracted course. The clinical spectrum of oral manifestations, particularly in secondary syphilis, is very broad, and the differential diagnosis should include aphthous ulcers, candidiasis, lichen planus, lupus erythematosus, non-specific erythema, erythroleukoplakia, squamous cell carcinoma and others. Oral syphilis must be included in the differential diagnosis of oral lesions in HIV-infected patients who should be under routine serological testing.
- ▶ Oral tuberculosis should also be regarded in the differential diagnosis of oral lesions in HIV-infected patients, particularly after the recent



FIGURE 208

HIV infection.  
Bacillary angiomatosis presenting  
as erythema and nodular swelling  
on the lower gingiva



FIGURE 209

HIV infection.  
Diffuse necrotizing ulceration  
on the upper gingiva due to  
*Pseudomonas aeruginosa*



dramatic increase of tuberculosis in this group of patients. Gingival involvement is rare and present as atypical, irregular ulceration.

- Some other oral infections caused by *Mycobacterium avium-intracellulare*, *Klebsiella pneumoniae*, *Enterobacter cloacae*, *Escherichia coli*, *Actinomyces israelii* and *Pseudomonas aeruginosa* have also been reported predominantly during the later stages of HIV disease. The lesions of all of these may develop on the gingiva (Fig. 209).

## Viral Infections

- ▶ The most frequent viral oral lesions in HIV-infected patients are caused by members of the herpesvirus group. Less frequent lesions may be caused by various types of human papilloma virus.

### Herpes Simplex Virus (see also p. 150)

- ▶ Primary and recurrent intraoral and perioral herpes simplex is a relatively frequent viral infection, with a prevalence of 5%–10%. The clinical features of oral herpesvirus infection are usually similar to those seen in non-HIV-infected patients; however, in HIV-infected patients with severe immune suppression (CD4 cell counts usually below 100 cells/mm<sup>3</sup>), widespread, severe, and prolonged lesions resemble primary herpetic gingivostomatitis (Fig. 210). In addition, recurrences are also common. Oral aciclovir (800–1200 mg/day) is usually the treatment of choice.

### Herpes Zoster (see also p. 169)

- ▶ Herpes zoster is one of the earliest clinical signs of HIV infection, particularly in individuals under 40 years of age with severe lesions, but without a history of immunosuppression. It would be more accurate to classify oral herpes zoster in young persons as a lesion „strongly associated with HIV infection“. In addition, HIV-infected individuals with herpes zoster develop AIDS more frequently than do matched controls, but the prevalence of oral herpes zoster is low (1%–2%). Gingival lesions may occur during second and third phase of trigeminal involvement (Fig. 211). The diagnosis of oral herpes zoster is based on clinical signs and symptoms similar to those seen in non-HIV-infected patients, with the exception of the severity of the lesions. Aciclovir, valaciclovir, or famciclovir is the treatment proposed.

### Cytomegalovirus

- ▶ Oral ulcerations associated with cytomegalovirus (CMV) infection have rarely been reported in HIV-infected patients. However, it is thought that oral lesions due to CMV may be more common, and thus the final diagnosis should be based on histological verification and molecular biology laboratory tests. These oral lesions are usually early signs of disseminated CMV infection and usually involved the gingiva (Fig. 212). The clinical features of CMV ulcers are similar to aphthous ulcers and other viral-associated ulcers as well as to non-specific ulcerations and drug-associated oral ulcerations usually seen in HIV-infected patients. Oral CMV infection is associated with severe immune suppression. Intravenous (IV) ganciclovir and aciclovir in high doses can be used for treatment.

FIGURE 210

HIV infection.  
Herpetic gingivitis



FIGURE 211

HIV infection.  
Herpes zoster on the upper gingiva  
and palate

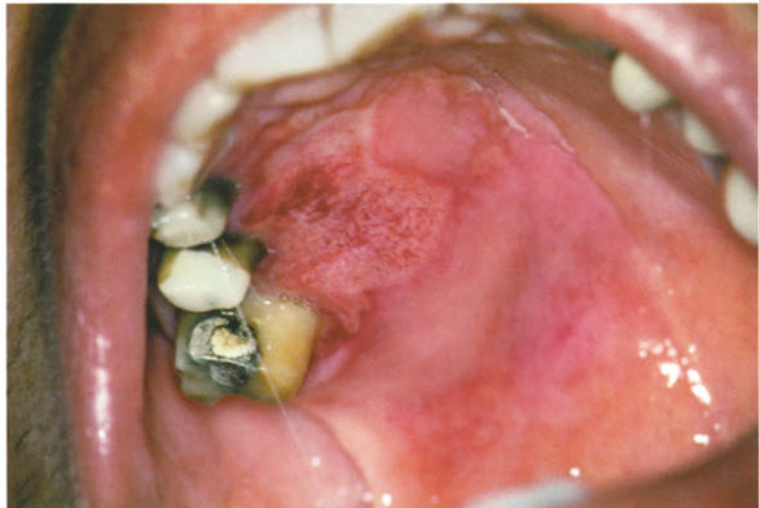


FIGURE 212

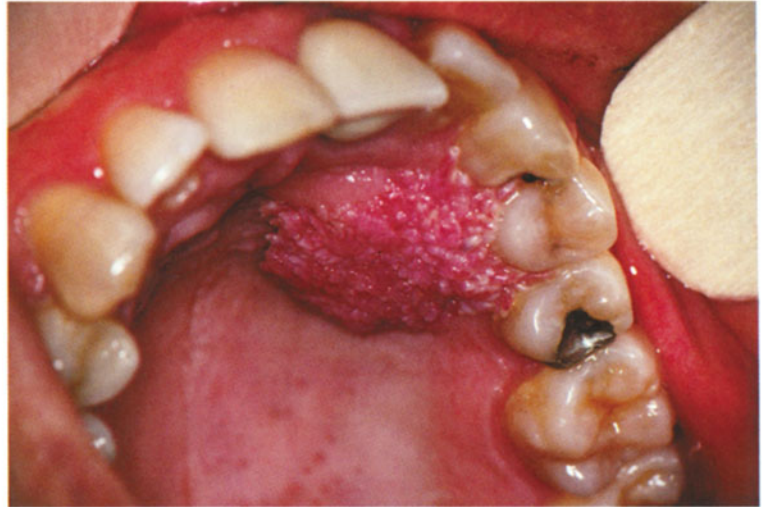
HIV infection.  
Gingival ulcer due to  
cytomegalovirus





FIGURE 213

HIV infection.  
Typical condyloma acuminatum  
on the upper gingiva



### Condyloma Acuminatum (see also p. 103)

- ▶ Condyloma acuminatum is relatively common in HIV-infected patients and the gingiva is the most common affected area (**Fig. 213**).

## Mycoses

### Linear Gingival Erythema

- ▶ Linear gingival erythema is a relatively uncommon gingival lesion presumably indicative of an underlying HIV infection. The lesion has received little attention in the literature and its exact prevalence rate in HIV-infected patients is not yet clear. Although the exact etiology remains unclear, a close association between oral candidiasis and linear gingival erythema exists.

#### Main Clinical Features

- ▶ Clinically, linear gingival erythema is characterized by a fiery red band along the margin of the gingiva and a punctate or diffuse erythema of the attached gingiva (**Figs. 214, 215**).
- ▶ There is no ulceration and evidence of pocket formation or attachment loss.
- ▶ Gingival bleeding may occur spontaneously or on probing. However, an important clinical sign of linear gingival erythema is now considered to be the absence of bleeding on probing.
- ▶ The lesion does not respond well to plaque-control measures or root planing and scaling. Plaque-control and a high level of oral hygiene are the treatments suggested. Persistent lesions may be related to candida infection and should be managed with antifungals.

FIGURE 214

HIV infection.  
Linear gingival erythema



FIGURE 215

HIV infection.  
Linear gingival erythema



### Candidiasis (see also p. 109)

- ▶ Oral candidiasis is the most common oral manifestation among patients with HIV, predominantly caused by *Candida albicans* and rarely by other *Candida* species. It may be the initial sign of HIV infection in patients who appear otherwise healthy. In addition, oral candidiasis is usually associated with an increased risk for progression to HIV disease. A significant interaction between oral candidiasis and CD4-cell-decline rate has also been detected. In one study of 700 HIV-infected patients, 70% with oral candidosis had CD4+ cell counts below 200 cells/mm<sup>3</sup>. The prevalence of the lesions ranges between 11% and 95%, depending on the study design and the patient group. Except for HIV infection, other predisposing, systemic and local factors can also influence the development of oral candidosis.
- ▶ In HIV-infected patients, candidal infection can occur in two major forms: pseudomembranous and erythematous. In addition, different forms of oral candidiasis can coexist in the same patient.



FIGURE 216

HIV infection.  
Pseudomembranous candidiasis  
on the upper gingiva



FIGURE 217

HIV infection.  
Severe pseudomembranous  
candidiasis on the upper gingiva



FIGURE 218

HIV infection.  
Erythematous candidiasis  
on the upper gingiva





- ▶ Pseudomembranous candidiasis presents as white or yellowish spots or plaques that can be wiped off, revealing an erythematous surface. The gingival lesions are not common (Fig. 216, 217). The soft palate, buccal mucosa, lateral borders of the tongue, commissures of lips are more frequently affected.
- ▶ Erythematous candidiasis presents as a red area usually on the palate and the dorsum of the tongue; less often gingival involvement may occur (Fig. 218). White spots and plaques may be seen, but these are not usually conspicuous.
- ▶ The treatment of oral candidiasis includes azole (ketoconazole, fluconazole, itraconazole) and polyene drugs (nystatin and amphotericin B). Resistance of oral candidosis to those drugs in HIV-infected patients is a common finding and recurrences are frequently seen.

### Systemic Mycoses (see also p. 193)

- ▶ Systemic or deep mycoses have, traditionally, been seen mainly in endemic areas and in developing countries, but they are now increasing in immunocompromised individuals, including HIV-infected patients. Gingival and oral lesions caused by systemic mycoses may occasionally be the primary or even the only manifestation, but they are usually associated with systemic signs and symptoms. HIV-infected patients are at particular risk from disseminating systemic mycoses.
- ▶ Of the systemic mycoses with oral manifestations, histoplasmosis and cryptococcosis have been most frequently encountered in HIV-infected patients.
- ▶ Sporadic cases of other systemic mycoses with oral manifestations have also been described in HIV-infected patients.
- ▶ Most of the systemic mycoses are diagnosed on the basis of a history of endemic areas, a recent trip abroad or an immunocompromised state. The laboratory diagnostic tests include smears, biopsy (staining with PAS, Gomori methenamine silver, Giemsa) and cultures from the affected tissues. In addition, physical examination, chest radiography and occasionally serological tests are necessary for the final diagnosis. In systemic mycoses, the oral lesions clinically present as ulcers, plaques or nodules and treatment includes systemic administration of azoles and amphotericin-B.

### Neoplasms

#### Kaposi's Sarcoma (see also p. 330)

- ▶ Kaposi's sarcoma (KS) is the most common HIV-associated neoplasm, occurring after new therapeutic regimens in approximately 5%–10% of the patients with acquired immunodeficiency syndrome (AIDS). It is a potentially multicentric neoplasm that may manifest only with regional involvement in the oropharynx or with multiple-site involvement, including viscera. Up to 60% of AIDS patients with Kaposi's sarcoma have oral lesions and usually the oral mucosa is the first site of involvement, with subsequent progress to multiple sites. The prevalence of KS increases significantly with the progression of the disease. It has been suggested that oral lesions of KS are usually associated with lower CD4 counts than those detected with cutaneous lesions alone. Intraoral KS has a higher predilection for homosexual or bisexual men compared to

FIGURE 219

AIDS.  
Early lesions of Kaposi's sarcoma  
presenting as red patch on  
the alveolar mucosa

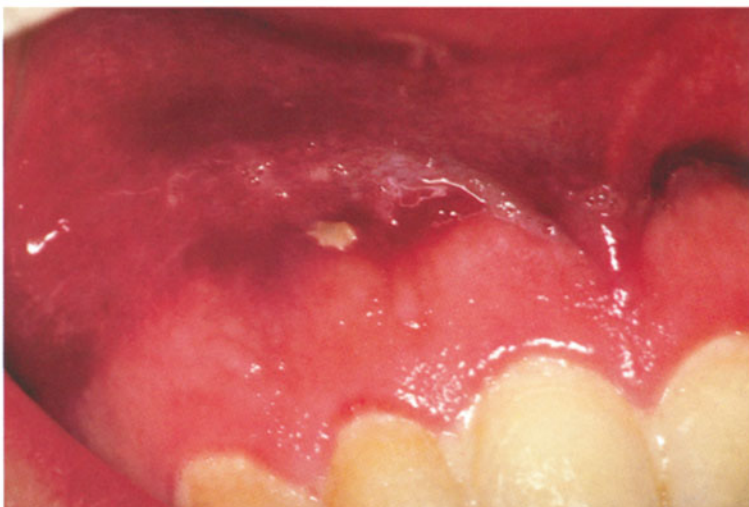


FIGURE 220

AIDS.  
Large Kaposi's sarcoma  
on the upper gingiva



other risk groups for HIV infection. Both the etiology and the pathogenesis of KS are still not well understood; however, human herpes virus 8 (HHV 8) plays an important role in the pathogenesis, presumably in association with other co-factors.

#### Main Clinical Features

- Clinically, in the early phase, the oral lesions appear as a red, purple, or pigmented macule, papule or patch. Later, solitary or multiple elevated and exophytic tumours with or without ulceration may be the most prominent clinical feature (Figs. 219, 220). The palate is the most common site of involvement, followed by the gingiva, buccal mucosa and tongue.



- ▶ A proposal has been suggested for clinical staging of oral KS depending on the extent (T) and the location (L) of tumour involvement and the clinical presentation or symptoms associated with the tumour (S). This staging might provide guidance for treatment and comparison of different treatment approaches among studies.
- ▶ A biopsy followed by histopathological examination of oral KS is necessary to confirm the clinical diagnosis.

#### Differential Diagnosis

- ▶ Bacillary angiomatosis
- ▶ Pyogenic granuloma
- ▶ Peripheral giant cell granuloma
- ▶ Leiomyoma
- ▶ Angiosarcoma
- ▶ Naevi
- ▶ Malignant melanoma
- ▶ Oral lesions of amyloidosis

#### Treatment

- ▶ Local and systemic treatment depending on the clinical stage. The local treatment can be surgical or laser excision, intralesional injection with vinblastine, sodium tetradecyl sulphate, or interferon and radiotherapy. Systemic treatment is single or combined chemotherapy (vincristine, vinblastine, bleomycin, Adriamycin).

### Non-Hodgkin's Lymphomas

- ▶ Non-Hodgkin's lymphoma is the second most common malignancy in HIV infection and it develops in patients with CD4 cell counts below 100 cells/mm<sup>3</sup>. Up to 3% of the patients would develop a non-Hodgkin lymphoma and the oral cavity accounts for 3% of all malignant lymphomas in patients with HIV infection. Not uncommonly, the mouth is the only site involved at the time of diagnosis. The posterior gingiva and fancer are frequently affected. All groups at risk for HIV infection are susceptible to the development of lymphoma, although some differences do exist in the clinical and histopathological features of individual groups. The majority of lymphomas are of B-cell origin, including the Burkitt's type. Rarely, Hodgkin's disease may manifest intra-orally and locate on the gingiva. Epstein-Barr virus is implicated as a cause for up to 50% of AIDS-associated lymphomas.

#### Main Clinical Features

- ▶ Clinically, oral lymphoma usually presents as a slowly or more often rapidly growing red or purplish, painless or painful mass that may be ulcerated (**Fig. 221**).
- ▶ Tooth loosening and paraesthesia are often the presenting symptoms.
- ▶ The posterior gingiva, the soft palate and fauces are the most commonly affected sites, while jaw involvement may also occur.
- ▶ The final diagnosis should be based on histopathological and histochemical findings.

#### Differential Diagnosis

- ▶ Kaposi's sarcoma
- ▶ Soft tissue and dental infection
- ▶ Systemic mycoses
- ▶ Salivary gland tumours

#### Treatment

- ▶ Treatment of oral lymphomas include multichemotherapy and radiation.



FIGURE 221

AIDS.  
Non-Hodgkin's lymphoma  
presenting as small ulcer  
on the upper gingiva



FIGURE 222

AIDS.  
Irregular ulcer on the gingival  
due to drug reaction (DDC)



### Drug Reactions

- ▶ As a result of treatment regimens, various drug-induced gingival and oral lesions commonly occur in HIV-infected patients (**Fig. 222**). Several of these reactions are associated with ulcerations, although the prevalence rate is generally unknown. This group of lesions includes stomatitis medicamentosa, erythema multiforme, Stevens-Johnson syndrome, toxic epidermal necrolysis, lichenoid reactions, oral pigmentation, oral ulcerations caused by DDC and foscarnet, oral lesions of agranulocytosis and face myositis.

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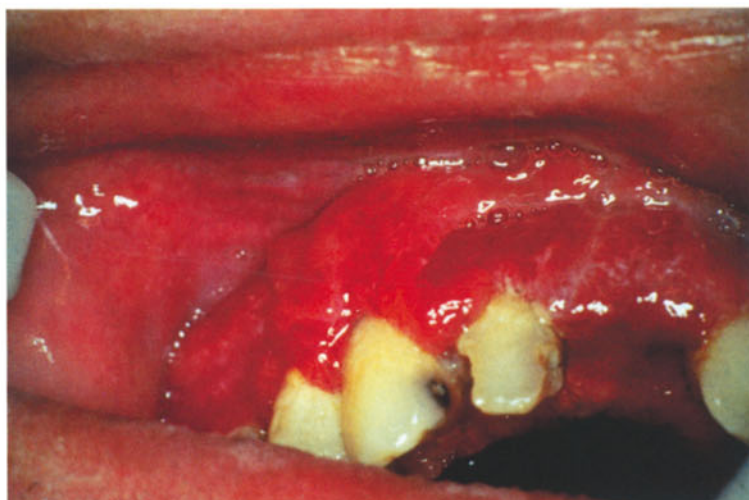
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### Histoplasmosis

- Definition** ▶ Histoplasmosis is a granulomatous systemic mycosis that is mainly endemic in the Ohio and Mississippi valleys in USA, in Latin America, India, east Asia and in Australia. However, sporadic cases may occur in other parts of the world, particularly in HIV infected patients.
- Etiology** ▶ *Histoplasma capsulatum* usually
- Classification** ▶ Three main forms are recognized:
- Primary acute pulmonary.
  - Chronic pulmonary.
  - Disseminated form.
- ▶ Oral lesions occur mainly in patients with disseminated disease and HIV disease, less frequently with pulmonary histoplasmosis.
- Gingival Involvement** ▶ Relatively common
- Other Involvement** ▶ Oral mucosa, relatively common
- ▶ Skin
  - ▶ Lung and other organs
- Main Clinical Features** ▶ The gingival lesions present as a chronic vegetating painful ulcer with a tendency to increase in size and less frequently as nodules (Fig. 223).

FIGURE 223

Histoplasmosis.  
Vegetating ulceration  
on the upper gingiva





- ▶ The oral mucosal lesions present also as painful ulcers or nodules usually on the tongue, palate and buccal mucosa.
- ▶ Oral lesions of histoplasmosis may be found in a patient without evidence of systemic histoplasmosis.
- ▶ Systemic manifestations include cough, fever, weight loss, chest pain, weakness, fatigue and involvements of lymph nodes, spleen, liver, kidneys, gastrointestinal tract and central nervous system.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Culture.
  - ▶ Serological tests.

- Differential Diagnosis**
- ▶ Oral tuberculosis
  - ▶ Squamous cell carcinoma
  - ▶ Other systemic mycoses
  - ▶ Eosinophilic ulcer
  - ▶ Wegener's granulomatosis
  - ▶ Non-Hodgkin's lymphoma

- Treatment**
- ▶ Fluconazole, itraconazole, amphotericin B

### Mucormycosis

- Definition**
- ▶ Mucormycosis or zygomycosis or phycomycosis is a relatively rare systemic mycosis that mainly affects immunocompromised patients (patients with leukaemia, non-Hodgkin's disease, severe, poorly controlled diabetes mellitus, AIDS, immunosuppressed patients).

- Etiology**
- ▶ Mainly *Mucor* and *Rhizopus* and rarely other species

- Gingival Involvement**
- ▶ Relatively common

- Other Involvement**
- ▶ Oral mucosa (palate)
  - ▶ Nasal cavity and paranasal sinuses
  - ▶ Lung, gastrointestinal tract
  - ▶ Cardiovascular and central nervous system

- Main Clinical Features**
- ▶ The oral manifestations occur in the rhinocerebral mucormycosis, which is the most common form of the disease.
  - ▶ The gingival lesions present as erythematous and oedematous areas, which soon progress in black necrotic ulcerations with severe alveolar bone destruction (**Fig. 224**).
  - ▶ The oral mucosal lesions usually appear as palatal, black necrotic ulcers, which quickly spread, creating extensive bone destruction.
  - ▶ The disease usually commences in the nasal cavity or paranasal sinuses with pain, nasal discharge and fever. Facial swelling, headache are also common.
  - ▶ Without proper treatment, mucormycosis may spread into the orbit and the brain, finally leading to death.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests and must be rendered early as soon as possible.
  - ▶ Biopsy and histopathological examination.
  - ▶ Smear examination.

FIGURE 224

Mucormycosis.  
Necrotic ulcerations  
and severe alveolar bone  
and palate destruction

**Differential Diagnosis**

- ▶ Severe necrotizing ulcerative gingivitis and stomatitis
- ▶ Malignant granuloma
- ▶ Wegener's granulomatosis
- ▶ Tuberculosis
- ▶ Non-Hodgkin's lymphoma
- ▶ Other systemic mycoses

**Treatment**

- ▶ Surgical debridement and systemic amphotericin B

**Paracoccidioidomycosis****Definition**

- ▶ Paracoccidioidomycosis, or South American blastomycosis, is a chronic granulomatous systemic mycosis which is endemic in Brazil and less common in Venezuela, Uruguay, Argentina and Colombia

**Etiology**

- ▶ *Paracoccidioides brasiliensis*

**Gingival Involvement**

- ▶ Relatively common

**Other Involvement**

- ▶ Oral mucosa, relatively common
- ▶ Lungs, always

**Main Clinical Features**

- ▶ The gingival lesions present as painful, chronic, granular vegetating ulcer with severe erythema, which may be localized or generalized (Fig. 225).
- ▶ The oral mucosal lesions also present as painful exophytic ulcerations with granular surface, usually occurring on the palate, tongue and lips.
- ▶ In several cases, the oral lesions follow tooth extraction.
- ▶ Chronic pulmonary disease with cough, dyspnea, hemoptysis, fever and weight loss are the presenting feature of the disease.

**Diagnosis**

- ▶ The clinical diagnosis should be confirmed by laboratory tests.
- ▶ Biopsy and histopathological examination.
- ▶ Smear examination or culture.
- ▶ Serological tests.

FIGURE 225

Paracoccidioidomycosis.  
Vegetating ulceration  
on the lower gingiva



- Differential Diagnosis**
- ▶ Pyostomatitis vegetans
  - ▶ Wegener's granulomatosis
  - ▶ Tuberculosis
  - ▶ Other systemic mycoses
- Treatment**
- ▶ Fluconazole, itraconazole
  - ▶ Amphotericin B
  - ▶ Sulfamethoxypyridazine

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## Haematological Disorders and Malignancies

### Familial Neutropenia

**Definition** ▶ Familial neutropenia is a rare haematological disorder characterized by quantitative decrease of neutrophils in the peripheral blood due to improper release of them from the bone marrow. An increased susceptibility of the patients to bacterial infection occurs.

**Etiology** ▶ It is inherited as an autosomal dominant and recessive trait.  
▶ Microbial dental plaque modified by quantitative leukocyte disorders are responsible for the periodontal lesions.

**Periodontal Involvement** ▶ Common  
▶ Periodontitis associated with haematological disorders

**Other Sites of Involvement** ▶ Oral mucosa  
▶ Skin, ear  
▶ Respiratory tract, urinary system

**Main Clinical Features** ▶ The main clinical manifestations are recurrent infections and are usually present at birth. Periodontal manifestations include fiery red oedematous gingivitis, severe periodontal lesions with deep periodontal pockets and extensive generalized bone loss leading to tooth mobility and loss (**Figs. 226, 227**).  
▶ Recurrent oral ulceration, which may lead to scar formation is common.

**FIGURE 226**

Familial neutropenia.  
Diffuse erythematous gingivitis  
on the upper gingiva



FIGURE 227

Familial neutropenia.  
Fiery red oedematous gingivitis



- ▶ Recurrent skin infections, otitis media, pneumonia, are constant manifestations.
- ▶ The severity of infections depend on quantitative neutrophil decrease.
- ▶ Affected children tend to improve with age and some undergo total remission in late childhood.

#### Diagnosis

- ▶ The clinical diagnosis should be confirmed by laboratory tests.
- ▶ Complete blood count.
- ▶ Radiographic examination.

#### Differential Diagnosis

- ▶ Aggressive periodontitis
- ▶ Cyclic neutropenia
- ▶ Agranulocytosis
- ▶ Leukaemia
- ▶ Chediak-Higashi syndrome
- ▶ Glycogen storage disease 1b
- ▶ Infantile genetic agranulocytosis
- ▶ Hypophosphatasia

#### Treatment

- ▶ Good oral hygiene.
- ▶ Periodontal treatment (scaling, surgery, antimicrobial agents).
- ▶ The systemic treatment must be left to the specialist haematologist.

### Cyclic Neutropenia

#### Definition

- ▶ Cyclic neutropenia is a rare haematological disorder, characterized by regular period depletion of neutrophil population, usually in 3-week cycles. The episode of neutropenia is usually short, persist for 5–10 days after which the neutrophil counts gradually return to normal.

#### Etiology

- ▶ The etiology remains unknown and the disorder is thought to be due to a defect of haematopoietic control. An autosomal dominant trait has been recorded in some cases.
- ▶ Microbial dental plaque modified by quantitative leukocyte disorders are responsible for the periodontal lesions.

**FIGURE 228**

Cyclic neutropenia.  
Diffuse gingival erythema  
and oedema



- Periodontal Involvement**
- ▶ Relatively common
  - ▶ Periodontitis associated with haematological disorders

- Other Sites of Involvement**
- ▶ Oral mucosa
  - ▶ Skin, ear

- Main Clinical Features**
- ▶ Periodontal manifestations include diffuse gingival inflammation and ulcerations, periodontal pockets and generalized bone loss (**Fig. 228**).
  - ▶ Painful, recurrent, oval oral ulcerations covered by a whitish membrane on an erythematous base usually occur.
  - ▶ Skin infections and otitis media may also occur during an episode of profound neutropenia.
  - ▶ Low-grade fever, malaise, headache, arthralgia, and cervical adenitis may occur.
  - ▶ The disease usually begins after 10 years of age.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Repeated complete blood count.
  - ▶ Bone marrow examination during periods of neutropenia shows decreased numbers of neutrophil precursors.

- Differential Diagnosis**
- ▶ Aggressive periodontitis
  - ▶ Familial neutropenia
  - ▶ Agranulocytosis
  - ▶ Chediak-Higashi syndrome
  - ▶ Glycogen storage disease 1b
  - ▶ Infantile genetic agranulocytosis
  - ▶ Leukaemia
  - ▶ Aphthous ulcers
  - ▶ Herpetic stomatitis

- Treatment**
- ▶ Good oral hygiene.
  - ▶ Periodontal treatment (scaling, surgery, antimicrobial agents).
  - ▶ The systemic treatment must be left to the specialist haematologist.



### Agranulocytosis

**Definition** ▶ Agranulocytosis is an acute haematological disorder characterized by severe depletion of neutrophils or all granulocytes in the blood.

**Etiology** ▶ It is often due to adverse drug reactions.  
▶ Idiopathic cases or cases induced by infections may also occur.  
▶ Microbial dental plaque modified by quantitative leukocyte disorders are responsible for the periodontal lesions.

**Periodontal Involvement** ▶ Common as generalized periodontal destruction  
▶ Periodontitis associated with haematological disorders

**Other Sites of Involvement** ▶ Oral mucosa (buccal mucosa, tongue, palate, tonsils)  
▶ Other mucosae (pharynx, larynx, oesophagus, gastrointestinal tract)  
▶ Respiratory tract

**Main Clinical Features** ▶ The periodontal lesions are early manifestations and are characterized by ulceration and necrosis of the marginal and attached gingiva associated with haemorrhage (**Fig. 229**). Severe generalized bone loss may also occur.  
▶ Necrotic ulcers covered by grey-white or dark, friable pseudomembranes are common findings on the oral mucosa. The ulcers do not have a red halo around them.  
▶ Halitosis, sialorrhoea, dysphagia, difficulty in swallowing, fever, malaise and headache are common.  
▶ Pulmonary and gastrointestinal infections are common.

**Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory test.  
▶ Complete blood count and bone marrow aspiration.

**Differential Diagnosis** ▶ Necrotizing ulcerative gingivitis and stomatitis  
▶ Familial neutropenia  
▶ Cyclic neutropenia  
▶ Acute leukaemia  
▶ Other types of neutropenia  
▶ Infectious mononucleosis  
▶ Aplastic anaemia

**Treatment** ▶ Good oral hygiene.  
▶ Periodontal treatment should follow the systemic treatment.  
▶ Systemic treatment must be left to the specialist haematologist.

### Aplastic Anaemia

**Definition** ▶ Aplastic anaemia is a severe haematological disorder characterized by acellular or markedly hypocellular bone marrow resulting in pancytopenia.

**Etiology** ▶ Drugs, chemicals, radiation, infections are usually associated with the disease. Idiopathic cases may also occur.

**Gingival Involvement** ▶ Common  
▶ Gingival lesions associated with blood dyscrasias

**FIGURE 229**

Agranulocytosis.  
Severe erythema and necrotic  
ulcerations of the gingiva

**FIGURE 230**

Aplastic anaemia.  
Early necrotic ulceration  
on the upper gingiva

**FIGURE 231**

Aplastic anaemia.  
Ecchymoses on the palate and tongue



- Other Sites of Involvement** ▶ Oral mucosa  
▶ Other mucosae, skin
- Main Clinical Feature** ▶ Gingival bleeding is a common early sign. Localized necrotic ulcerations on the marginal gingiva and the interdental papillae, mimic necrotizing ulcerative gingivitis (**Fig. 230**).  
▶ The oral mucosal lesions appear as necrotic ulcers, petechiae, ecchymoses and pallor of the oral mucosa (**Fig. 231**).  
▶ Pallor, petechiae, or ecchymoses and purpuric spots of the skin and other mucous membranes are also early signs.  
▶ The onset of the disease is usually insidious and the most common symptoms are fever, weakness, fatigue, fever and mild haemorrhage.  
▶ The course of the disease depends on the severity of aplasia.
- Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.  
▶ Complete blood count and bone marrow aspiration and biopsy.
- Differential Diagnosis** ▶ Necrotizing ulcerative gingivitis and stomatitis  
▶ Vitamin C deficiency  
▶ Agranulocytosis  
▶ Neutropenia  
▶ Leukaemia  
▶ Thrombocytopenic purpura  
▶ Infectious mononucleosis
- Treatment** ▶ Good oral hygiene.  
▶ Topical oral antiseptics.  
▶ Systemic treatment must be left to the specialist haematologist.

### Thrombocytopenic Purpura

- Definition** ▶ Thrombocytopenic purpura is a haematological disorder characterized by a profound decrease in platelets in the peripheral blood.
- Etiology** ▶ It may be idiopathic due to a primary failure of the bone marrow to generate platelets or secondary due to a myelotoxic drug, virus and HIV infections, leukaemias and other autoimmune diseases, malignancies, radiation, etc.
- Gingival Involvement** ▶ Common
- Other Sites of Involvement** ▶ Other mucosae (conjunctiva, nose, gastrointestinal, urinary)  
▶ Skin
- Main Clinical Features** ▶ Gingival bleeding, spontaneous and/ or prolonged is a constant early feature (**Fig. 232**). There is no evidence that thrombocytopenic purpura increases susceptibility to periodontal disease.  
▶ On the oral mucosa' petechiae, ecchymoses and haematomas may occur.  
▶ Bleeding episodes from gastrointestinal and urinary tracts and epistaxis are common.  
▶ Purpuric skin rash and occasionally bleeding occur.  
▶ The breadth and the severity of the manifestations are related to the extent of platelet reduction.



FIGURE 232

Thrombocytopenic purpura.  
Gingival bleeding



- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Complete blood count and bone marrow aspiration.

- Differential Diagnosis**
- ▶ Leukaemia
  - ▶ Aplastic anaemia
  - ▶ Agranulocytosis

- Treatment**
- ▶ Good oral hygiene.
  - ▶ Systemic and etiological treatment must be left to the specialist haematologist.

### Plasminogen Deficiency

- Definition**
- ▶ Plasminogen deficiency, or hypoplasminogenaemia, is a deficiency in plasminogen. In health, body fluid fibrinolytic activity clears fibrin deposits, but if plasminogen is deficient this mechanism fails, with fibrin deposition occurring. Perhaps surprisingly, there is rarely a tendency to thrombosis.

- Etiology**
- ▶ An autosomal recessive disorder

- Gingival Involvement**
- ▶ Common

- Other Sites of Involvement**
- ▶ Corneal involvement and blindness
  - ▶ Congenital occlusive hydrocephalus

- Main Clinical Features**
- ▶ Generalized painful ulcerated gingival swelling since late childhood (**Fig. 233**).
  - ▶ Gingival enlargement is more severe in interdental papillae (**Fig. 234**).

- Diagnosis**
- ▶ The clinical diagnosis should always be confirmed by laboratory tests.
  - ▶ Histopathological examination.
  - ▶ Plasminogen concentration.

FIGURE 233

Plasminogen deficiency.  
Gingival ulcerations



FIGURE 234

Plasminogen deficiency.  
Severe gingival swelling  
and minor ulcerations



- Differential Diagnosis**
- ▶ Hereditary gingival fibromatosis and related disorders
  - ▶ Lipoid proteinosis
  - ▶ Mucocutaneous amyloidosis
  - ▶ Mucopolysaccharidosis I-H

- Treatment**
- ▶ Topical heparin or intravenous purified plasminogen concentrate

### Myelodysplastic Syndrome

- Definition**
- ▶ Myelodysplastic syndromes form a heterogeneous group of refractory anaemias often associated with thrombocytopenia, neutropenia, and/or monocytosis. Older individuals are more often affected.
- Etiology**
- ▶ The exact etiology remains unknown. It may develop secondary to radiotherapy and chemotherapy.

**FIGURE 235**

Myelodysplastic syndrome.  
Gingival swelling  
and early ulcerations

**FIGURE 236**

Myelodysplastic syndrome.  
Gingival ulcerations



**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ Oral mucosa  
▶ Other mucosae  
▶ Skin and other systems

**Main Clinical Features** ▶ The gingival lesions may appear as chronic localized swelling and/or ulcerations, associated with bleeding (**Figs. 235, 236**).  
▶ The oral mucosal lesions appear as persistent and recurrent painful ulcerations and bleedings. Less often oral candidiasis may develop.  
▶ Multiple bacterial infections and haemorrhage are the main features of the disease due to neutropenia and thrombocytopenia.  
▶ Malaise, fatigue, fever are common symptoms.

**Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.  
▶ Complete blood count and bone marrow aspiration and biopsy.



- Differential Diagnosis**
- ▶ Leukaemias
  - ▶ Agranulocytosis
  - ▶ Neutropenia
  - ▶ Aplastic anaemia
  - ▶ Thrombocytopenia
  - ▶ Non-Hodgkin lymphoma
  - ▶ Multiple myeloma
  - ▶ Langerhans cell histiocytosis

- Treatment**
- ▶ Good oral hygiene.
  - ▶ Topical oral antiseptics.
  - ▶ Systemic treatment must be left to the specialist haematologist.

### Leukaemias

- Definition**
- ▶ Leukaemias are an heterogenous group of serious malignant disorders characterized by defects of leukocyte cell maturation and proliferation

- Etiology**
- ▶ The etiology remains obscure, although several factors are implicated such as genetic, viral infections, chemicals, radiation, immunodeficiency.

- Classification**
- ▶ Depending on the cell type primarily affected, leukaemias are classified into:
    - Lymphocytic
      - Acute
      - Chronic
    - Non-lymphocytic
      - Acute
      - Chronic

- Gingival Involvement**
- ▶ Common. Approximately 30 % of patients with acute and 10 % with chronic leukaemias present with gingival lesions.
  - ▶ Pronounced inflammatory reaction of gingiva in relation to the dental plaque present. However, dental plaque is not a prerequisite for gingival lesions.

- Other Sites of Involvement**
- ▶ Oral mucosa
  - ▶ Skin
  - ▶ Lymph nodes, spleen, liver

- Main Clinical Features**
- ▶ Generalized gingival swelling of variable severity may appear usually in acute and rarely in chronic leukaemia (Figs. 237–239). Severe gingival enlargement is usually a feature of acute monocytic leukaemia (Figs. 240–243). The gingival enlargement initially starts at the interdental papilla and is followed by the marginal and attached gingiva. The enlargement may be due to both plaque-related chronic inflammation and to infiltration by leukaemic cells.
  - ▶ Gingival bleeding may be one of the initial oral signs in the:
    - Acute form
    - Chronic form
  - ▶ Rarely, rapid alveolar bone loss may occur, particularly in the patient with pre-existing periodontitis. Not infrequently, gingival lesions are the initial manifestation of leukaemia.
  - ▶ Non-specific ulcerations of the oral mucosa and bleeding may occur.

FIGURE 237

Acute myeloblastic leukaemia.  
Early gingival swelling



FIGURE 238

Acute myeloblastic leukaemia.  
Severe gingival enlargement



FIGURE 239

Acute myeloblastic leukaemia.  
Severe gingival enlargement  
and bleeding





FIGURE 240

Acute myelomonocytic leukaemia.  
Early red gingival swelling



FIGURE 241

Acute myelomonocytic leukaemia.  
Severe gingival enlargement



FIGURE 242

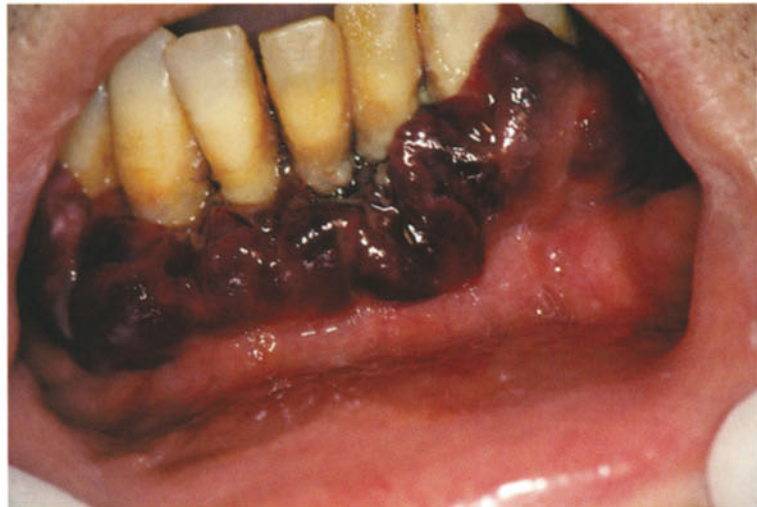
Acute myelomonocytic leukaemia.  
Severe gingival enlargement





**FIGURE 243**

Acute monocytic leukaemia.  
Severe red gingival swelling as an  
initial sign of the leukaemia



- ▶ Early symptoms are pallor, anorexia, irritability, fatigability, low fever, bleeding and lymph node enlargement.
- ▶ During chemotherapy, oral viral, bacterial and candida infections are common.

**Diagnosis**

- ▶ The clinical diagnosis should be confirmed by laboratory tests.
- ▶ Complete blood count and bone marrow biopsy.
- ▶ Biopsy of extramedullary tissue involved.

**Differential Diagnosis**

- ▶ Necrotizing ulcerative gingivitis
- ▶ Aplastic anaemia
- ▶ Agranulocytosis
- ▶ Neutropenia
- ▶ Multiple myeloma
- ▶ Langerhans cell histiocytosis
- ▶ Myelodysplastic syndrome

**Treatment**

- ▶ Good oral hygiene.
- ▶ Topical oral antiseptics.
- ▶ Systemic treatment must be left to the specialist haematologist.

**Multiple Myeloma****Definition**

- ▶ Multiple myeloma is a relatively rare malignant plasma cell disorder due to overproduction of specific immunoglobulins, not directed against any recognized epitope.

**Etiology**

- ▶ Unknown

**Gingival Involvement**

- ▶ Relatively rare

**Other Sites of Involvement**

- ▶ Oral mucosa (about 14%)
- ▶ Skin
- ▶ Skull, skeleton, including jaws
- ▶ Kidneys

FIGURE 244

Multiple myeloma.  
Localized gingival swelling



FIGURE 245

Multiple myeloma.  
Localized severe gingival swelling



- Main Clinical Features**
- ▶ Gingival bleeding, localized or generalized gingival swelling and gingival necrotic ulceration usually associated with alveolar bone destruction are the hallmarks of the neoplastic process (Figs. 244, 245). Gingival paraesthesia or anaesthesia may also occur.
  - ▶ Alveolar bone pain, swelling and tooth mobility are also common.
  - ▶ Bone pain is usually the presenting symptom. Fatigue, fever, petechiae of the skin, amyloid deposits in various soft tissues may occur.
  - ▶ The disease usually involves individuals over 50 years of age.
- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory test.
  - ▶ Biopsy and histopathological examination of the lesional tissue.
  - ▶ Radiographic examination.
  - ▶ Serum and urine protein electrophoresis.
- Differential Diagnosis**
- ▶ Necrotizing ulcerative gingivitis
  - ▶ Leukaemia
  - ▶ Aplastic anaemia

- ▶ Lymphomas
- ▶ Neutropenia
- ▶ Agranulocytosis
- ▶ Langerhans cell histiocytosis

- Treatment**
- ▶ Good oral hygiene.
  - ▶ Topical oral antiseptics.
  - ▶ Periodontal treatment.
  - ▶ Systemic treatment must be left to the specialist.

### Soft Tissue Plasmacytoma

- Definition**
- ▶ Soft tissue plasmacytoma, or solitary extramedullary plasmacytoma, is an unusual monoclonal neoplasm that consists of plasma cells indistinguishable from those seen in multiple myeloma.

- Etiology**
- ▶ Unknown

- Gingival Involvement**
- ▶ Rare

- Other Sites of Involvement**
- ▶ Oral mucosa (palate, tongue, floor of the mouth)
  - ▶ Nasopharynx, nasal cavity, paranasal sinuses, tonsils

- Main Clinical Features**
- ▶ Localized non-tender, well-circumscribed gingival swelling is the most common clinical feature. The gingival surface is usually normal or slightly red and rarely may be ulcerated (**Fig. 246**). Periodontal destruction may rarely occur. The size of the swelling varies from one several centimetres in diameter.
  - ▶ Similar clinical features present at the other sites of involvement.
  - ▶ A number of patients with primary soft tissue plasmacytoma may ultimately develop multiple myeloma.
  - ▶ The disease affects more often males over 50 years of age.

- Diagnosis**
- ▶ The clinical diagnosis should always be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Immunohistochemical examination for H and L chain.

**FIGURE 246**

Soft tissue plasmacytoma.  
Localized gingival swelling





- Differential Diagnosis**
- ▶ Non-Hodgkin's lymphoma
  - ▶ Multiple myeloma
  - ▶ Dental and periodontal abscesses
  - ▶ Gingival cyst of adults
  - ▶ Pleomorphic adenoma

- Treatment**
- ▶ Surgical excision usually in combination with radiotherapy, chemotherapy

### Non-Hodgkin's Lymphoma

- Definition**
- ▶ Non-Hodgkin's lymphoma is a heterogeneous group of neoplastic disorders that encompass neoplastic proliferation of lymphocytic cell lines (B- and T-cell origin)

- Etiology**
- ▶ Transforming genes and viruses may be involved in the pathogenesis of some forms of lymphomas.

- Gingival Involvement**
- ▶ Rare. The posterior gingiva and retromolar area are more often involved.

- Other Sites of Involvement**
- ▶ Oral mucosa (soft palate, base of the tongue, buccal mucosa) is rarely involved.
  - ▶ Abdominal, head and neck and mediastinal lymph nodes and Waldeyer's ring are the most common sites of involvement.
  - ▶ Extranodal involvement is rare.

- Main Clinical Features**
- ▶ Localized, painless, soft gingival swelling, occasionally associated with ulceration are the hallmarks of the gingival involvement (**Figs. 247, 248**). Periodontal destruction and bone loss are unusual.
  - ▶ The oral mucosa lesions appear as a soft, painless, reddish or purplish diffuse swellings. The lesion may be ulcerated.
  - ▶ In the head and neck area the disease presents as a painless mass of lymph nodes that increase slowly. As the disease progresses, the enlarged nodes become more numerous and fixed to surrounding tissues.
  - ▶ Fever, night sweats, weight loss may occur.

- Diagnosis**
- ▶ The clinical diagnosis should always be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Immunohistochemical examination.
  - ▶ Radiography and CT.

- Differential Diagnosis**
- ▶ Soft tissue plasmacytoma
  - ▶ Hodgkin's disease
  - ▶ Leukaemias
  - ▶ Multiple myeloma
  - ▶ Myelodysplastic syndrome
  - ▶ Dental and periodontal abscesses

- Treatment**
- ▶ Chemotherapy
  - ▶ Radiotherapy

FIGURE 247

Non-Hodgkin's lymphoma.  
Localized gingival swelling  
with ulceration



FIGURE 248

Non-Hodgkin's lymphoma.  
Gingival and palatal swelling  
(the ulcer is due to the biopsy)



### Burkitt's Lymphoma

**Definition** ▶ Burkitt's lymphoma is a malignant B-cell lymphoma arising within germinal cells of the lymph nodes. The disease is rare in Western countries and common in Africa, usually presenting between 10 and 12 years of age.

**Etiology** ▶ Epstein-Barr virus.  
▶ Immunodeficiencies, AIDS, autoimmune disease may predispose to the development of the lymphoma.

**Gingival Involvement** ▶ Very rare

**Other Sites of Involvement** ▶ Oral mucosa, rare.  
▶ Jaws, very often (60%–70%). The maxilla is twice as often affected as the mandible.  
▶ Bone marrow, abdominal region and central nervous system may be involved.

FIGURE 249

Burkitt's lymphoma.  
Localized soft tumour on the gingiva



- Main Clinical Features**
- ▶ The gingival lesions appear as localized gingival swelling or exophytic soft mass tumour that may be ulcerated (**Fig. 249**). The colour of the mass is reddish. Tumour growth is usually rapid and may produce periodontal destruction and tooth mobility.
  - ▶ Rapidly proliferative swelling of the jaws, which may produce marked destruction of the facial bone and facial nerve paresis, are the main clinical features of the disease. Bone pain, tenderness and paraesthesia may occur.
  - ▶ The prognosis depends on the stage of the disease.
- Diagnosis**
- ▶ The clinical diagnosis should always be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Immunohistochemical examination.
  - ▶ Panoramic radiographic examination, CT.
- Differential Diagnosis**
- ▶ Kaposi's sarcoma
  - ▶ Peripheral giant cell granuloma
  - ▶ Odontogenic tumours
  - ▶ Ewing's sarcoma
  - ▶ Non-Hodgkin's lymphomas (other types)
  - ▶ Amyloidosis
  - ▶ Fibrous dysplasia
  - ▶ Cherubism
- Treatment**
- ▶ Radiotherapy and chemotherapy
  - ▶ Surgical excision

### Cutaneous T-cell Lymphoma

- Definition**
- ▶ Cutaneous T-cell lymphoma, or mycosis fungoides, is a rare, slowly progressive non-Hodgkin's lymphoma primarily involving the skin.
- Etiology**
- ▶ It remains obscure.
- Gingival Involvement**
- ▶ Rare



**Other Sites of Involvement**

- ▶ Oral mucosa, rare
- ▶ Skin, always
- ▶ Visceral

**Main Clinical Features**

- ▶ Three clinical stages are recognized:
  - Erythematous.
  - Plaque.
  - Tumour.
- ▶ The gingival involvement usually occurs during the plaque and tumour stage and always after the skin lesions. Localized painful ulcers and, less often, red plaques or soft masses are the most common gingival lesions (**Fig. 250**).
- ▶ The oral mucosa lesions present as non-specific erythematous plaques or superficial ulcerations or ulcerated tumours.
- ▶ The skin lesions begin as an intensely pruritic eruption followed after a long time by the indurated plaque stage and finally by the tumour stage (**Fig. 251**).
- ▶ Males are more frequently affected, usually between 55 and 66 years of age.

**FIGURE 250**

Cutaneous T-cell lymphoma.  
Irregular ulceration on the  
upper gingiva

**FIGURE 251**

Cutaneous T-cell lymphoma.  
Perioral skin lesions



- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Immunohistochemical examination.

- Differential Diagnosis**
- ▶ Non-Hodgkin's lymphoma
  - ▶ Leukaemia
  - ▶ Lupus erythematosus

- Treatment**
- ▶ Good oral hygiene
  - ▶ Photochemotherapy, chemotherapy and radiation for the skin lesions

### Hodgkin's Disease

- Definition**
- ▶ Hodgkin's disease is a malignant lymphoproliferative disorder.

- Etiology**
- ▶ Genetic factors, viruses and environmental factors and AIDS, may be involved in the pathogenesis.

- Gingival Involvement**
- ▶ Very rare

- Other Sites of Involvement**
- ▶ Oral mucosa, very rare.
  - ▶ Lymph nodes, always.
  - ▶ Extranodular involvement may rarely occur.

- Main Clinical Features**
- ▶ Localized gingival soft swelling or non-specific ulceration are the presenting signs on the gingiva (**Fig. 252**)
  - ▶ The most frequent presenting feature is cervical lymph node (single or multiple) enlargement. The involved lymph nodes are firm, mobile and usually non-tender. Low-grade fever, night sweats, weight loss, anorexia, fatigue, malaise, weakness are common symptoms.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Immunohistochemical examination.
  - ▶ Radiography and CT.

- Differential Diagnosis**
- ▶ Non-Hodgkin's lymphoma
  - ▶ Leukaemias
  - ▶ Plasmacytoma

- Treatment**
- ▶ Good oral hygiene
  - ▶ Radiotherapy, chemotherapy
  - ▶ Bone marrow transplantation

FIGURE 252

Hodgkin's disease.  
Localized ulceration on the gingiva  
in a patient with AIDS



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### Erythema Multiforme

**Definition** ▶ Erythema multiforme is an acute mucocutaneous disease that occurs most frequently in young males.

**Etiology** ▶ Several factors have been implicated in the etiology of erythema multiforme, including drugs (sulphonamides, penicillin, barbiturates, non-steroidal anti-inflammatory drugs, phenytoin and others), herpes simplex virus infection, *Mycoplasma pneumoniae* infection. However, in more than 50% of the cases, a causative factor cannot be found.

▶ Two main forms of the disease are recognized:

- Erythema multiforme minor features distinct skin lesions with or without mucosal involvement.
- Erythema multiforme major, or Stevens-Johnson syndrome, which almost invariably involves the oral and other mucosae and the skin as well.

**Gingival Involvement** ▶ Relatively uncommon

▶ Non-plaque-induced lesions with no attachment loss

**Other Sites of Involvement** ▶ Oral mucosa (lips, buccal mucosa, tongue, palate), about 20%–30% in the minor form and almost invariably there in the major form of the disease.

▶ Other mucosae (conjunctiva, genitalia).

▶ Skin is very often affected in a symmetrical pattern (palms, soles, backs of the hands and feet, neck, trunk).

**Main Clinical Features** ▶ The gingival lesions may appear as diffuse erythema mimic desquamative gingivitis or localized or diffuse painful erosions covered by a whitish pseudomembrane (Figs. 253, 254).

▶ The oral mucosa lesions appear as small vesicles that rupture, creating extensive painful erosions covered by pseudomembranes and haemorrhagic crusts (Fig. 255).

▶ Photophobia, conjunctivitis, keratitis, uveitis and panophthalmitis may also occur.

▶ The skin lesions present as erythematous maculopapules, usually creating the characteristic target or iris lesions (Fig. 256). Less often purpuric plaques and bullous formation may appear, usually in association with target lesions.

▶ The disease recurs in 20%–30% of the cases.

FIGURE 253

Erythema multiforme.  
Early lesions on the gingiva  
presenting as desquamative gingivitis



FIGURE 254

Erythema multiforme.  
Erythema and erosions of the gingiva  
mimic desquamative gingivitis



- Diagnosis**
- ▶ The diagnosis is usually based on clinical criteria. However, occasionally laboratory test should be done.
  - ▶ Biopsy and histopathological examination.
  - ▶ Direct immunofluorescence.
- Differential Diagnosis**
- ▶ Primary herpetic gingivostomatitis
  - ▶ Herpetiform ulcers
  - ▶ Cicatricial pemphigoid
  - ▶ Bullous pemphigoid
  - ▶ Linear IgA disease
  - ▶ Pemphigoid gestations
  - ▶ Pemphigus
- Treatment**
- ▶ Systemic corticosteroids
  - ▶ Aciclovir or other new antivirals may be helpful in the management of patients with recurrent erythema multiforme

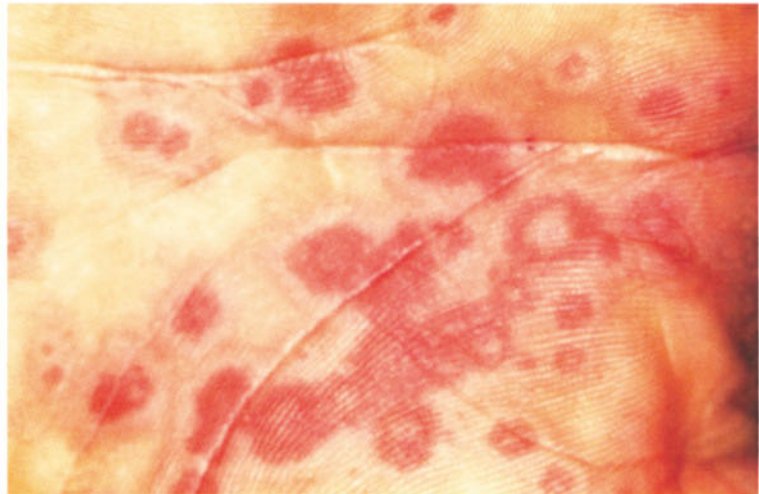


**FIGURE 255**

Erythema multiforme.  
Extensive erosions on the tongue  
and lower lip

**FIGURE 256**

Erythema multiforme.  
Characteristic target-like lesions  
on the palm



### Toxic Epidermal Necrolysis

**Definition** ▶ Toxic epidermal necrolysis or Lyell's disease is a rare exfoliative mucocutaneous disease with high mortality.

**Etiology** ▶ Drug-induced reaction (sulphonamides, antibiotics, non-steroidal anti-inflammatory agents, anticonvulsants)

**Gingival Involvement** ▶ Common  
▶ Non-plaque-induced lesions with no attachment loss

**Other Sites of Involvement** ▶ Oral mucosa (lips, buccal mucosa, tongue, palate)  
▶ Other mucosae (conjunctiva, oropharynx, genitalia, anus)  
▶ Skin (entire skin surface may be involved)

**Main Clinical Features** ▶ The disease usually commences with a cough, sore throat, burning eyes, low fever, malaise, followed after 1–2 days by skin and mucous membrane lesions.

FIGURE 257

Toxic epidermal necrolysis.  
Early lesions presenting as erythema  
and erosions on the gingiva mimic  
necrotizing ulcerative gingivitis



FIGURE 258

Toxic epidermal necrolysis.  
Sheet-like loss of the epidermis



- ▶ The gingival lesions appear as dense inflammation, with blister formation leading to painful widespread erosions (Fig. 257).
- ▶ The oral lesions are characterized by diffuse erythema, blistering and painful, widespread erosions.
- ▶ The skin lesions appear as a sheet-like loss of epidermis (Fig. 258). The epidermis is raised by flaccid blisters, which spread on pressure. Nikolsky's sign is positive.

#### Diagnosis

- ▶ The diagnosis is usually made on clinical level.
- ▶ Biopsy and histopathological examination.

#### Differential Diagnosis

- ▶ Erythema multiforme major
- ▶ Pemphigus
- ▶ Staphylococcal scalded skin syndrome

#### Treatment

- ▶ Patients must be admitted as soon as possible to an intensive care unit for management.

### Cicatricial Pemphigoid

- Definition** ▶ Cicatricial pemphigoid or benign mucous membrane pemphigoid is a chronic bullous disease that primarily affects mucous membranes and rarely the skin.
- ▶ The disease affects more frequently females than males (1.5:1), usually during the 5th to 6th decade of life.
- Etiology** ▶ Autoimmune
- Gingival Involvement** ▶ Very common, approximately 60 %
- ▶ Non-plaque-induced lesions with no attachment loss
- Other Sites of Involvement** ▶ Oral mucosa (soft palate, buccal mucosa, tongue, lips), 100 %
- ▶ Other mucosae (ocular, nose, genitalia, pharynx, larynx, anus)
- ▶ Skin, very rare (5 %)
- Main Clinical Features** ▶ Frequently the disease affects exclusively the gingiva in the form of desquamative gingivitis (erythema, oedema, desquamation of the epithelium or bullous formation after pressure of the gingiva) (Figs. 259–261).
- ▶ The oral mucosa lesions present as erythema and bullae formation that rupture quickly, leaving painful erosions (Fig. 262).
- ▶ The lesions usually recur and persist for a long time, leading occasionally to atrophy or scarring (except gingiva).
- ▶ The ocular lesions consist of conjunctivitis, symblepharon, trichiasis, dryness and opacity of the cornea, occasionally leading to complete blindness (Fig. 263).
- Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.
- ▶ Biopsy and histopathological examination.
- ▶ Direct immunofluorescence.
- ▶ Indirect immunofluorescence.
- Differential Diagnosis** ▶ Lichen planus
- ▶ Bullous pemphigoid
- ▶ Pemphigus
- ▶ Linear IgA disease
- ▶ Epidermolysis bullosa acquisita
- ▶ Chronic ulcerative stomatitis
- ▶ Plasma cell gingivitis
- ▶ Herpetic gingivostomatitis
- Treatment** ▶ Corticosteroid topical or systemic depending on disease activity
- ▶ Immunosuppressive treatment (azathioprine, ciclosporin) only in severe and persistent cases, always as adjuvant to corticosteroid therapy



**FIGURE 259**

Cicatricial pemphigoid.  
Early lesion on the gingiva  
presenting as small haemorrhagic  
bulla formation

**FIGURE 260**

Cicatricial pemphigoid.  
Typical desquamative gingivitis

**FIGURE 261**

Cicatricial pemphigoid.  
Localized lesions on the gingiva in  
the form of desquamative gingivitis



FIGURE 262

Cicatricial pemphigoid.  
Extensive erosions on the palate

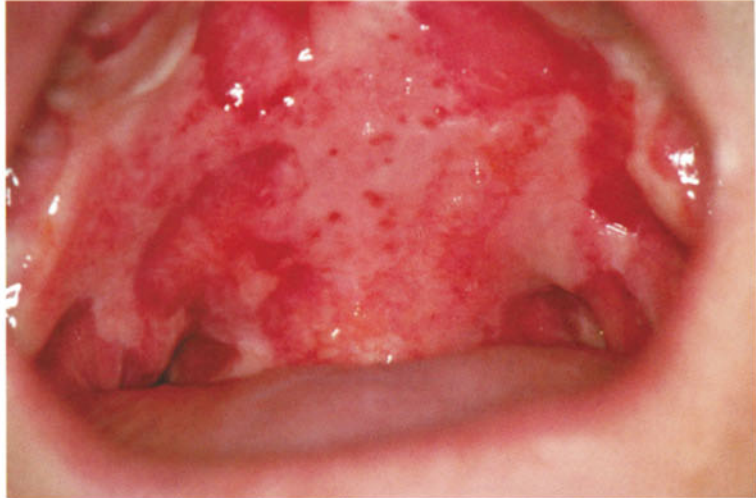


FIGURE 263

Cicatricial pemphigoid.  
Conjunctivitis and symblepharon



### Bullous Pemphigoid

- Definition**
- ▶ Bullous pemphigoid is a chronic bullous disease that primarily affects the skin and less often the oral mucosa (30%–40%).
  - ▶ The disease affects more frequently females than males (1.7:1), usually during the 5th to 6th decade of life.

- Etiology**
- ▶ Autoimmune

- Gingival Involvement**
- ▶ Relatively common, about 10%–15%
  - ▶ Non-plaque-induced lesions with no attachment loss

- Other Sites of Involvement**
- ▶ Oral mucosa (palate, buccal mucosa, tongue, lips), about 30%–40%
  - ▶ Other mucosae (conjunctiva, oesophagus, vagina, anus)
  - ▶ Skin, always



FIGURE 264

Bullous pemphigoid.  
Slight erythema of the gingiva and  
small haemorrhagic bulla formation



FIGURE 265

Bullous pemphigoid.  
Multiple small haemorrhagic bullae  
on the gingiva



FIGURE 266

Bullous pemphigoid.  
Small bullae on the skin





- Main Clinical Features**
- ▶ Gingival involvement usually appears in the form of desquamative gingivitis (erythema, oedema, desquamation of the epithelium) or as a localized bullous formation that ruptures quickly, leaving painful erosions (Figs. 264, 265).
  - ▶ Localized or widespread lesions may also appear in other oral mucosa sites.
  - ▶ The skin lesions usually begin as a non-specific generalized rash and ultimately tense bullae develop isolated or in clusters that rupture, leaving erosions without a tendency to extend peripherally (Fig. 266).
  - ▶ Nikolsky's sign is negative.
  - ▶ The disease has a chronic course with exacerbations and remissions and ultimately good prognosis.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Direct immunofluorescence.

- Differential Diagnosis**
- ▶ Cicatricial pemphigoid
  - ▶ Linear IgA disease
  - ▶ Pemphigoid gestationis
  - ▶ Epidermolysis bullosa acquisita
  - ▶ Pemphigus
  - ▶ Plasma cell gingivitis

- Treatment**
- ▶ Corticosteroid topical and/or systemic
  - ▶ Immunosuppressives as adjuvant therapy

### Linear IgA Disease

- Definition**
- ▶ Linear IgA disease is a rare vesiculobullous mucocutaneous disorder characterized by linear homogeneous deposition of IgA along the dermoepidermal junction on direct immunofluorescence testing.
  - ▶ The disease is more common in females than males, with an average onset between 40 and 50 years of age. Children may also be affected.

- Etiology**
- ▶ Autoimmune

- Gingival Involvement**
- ▶ Rare, approximately 5%–10%
  - ▶ Non-plaque-induced lesions with no attachment loss

- Other Sites of Involvement**
- ▶ Oral mucosa (about 20%–25%)
  - ▶ Other mucosae (conjunctiva, nasal, larynx) rarely
  - ▶ Skin, common, in a symmetrical and cluster form distribution (perioral, genital, buttocks, arms)

- Main Clinical Features**
- ▶ Gingival involvement may appear either in the form of desquamative gingivitis or as a localized bullous formations that rupture, leaving quickly non-specific painful erosions (Figs. 267, 268).
  - ▶ Localized or widespread lesions may also develop in other oral mucosa sites.
  - ▶ Skin lesions are characterized by pruritus and vesicles distributed in a symmetrical pattern (Fig. 269).
  - ▶ The disease has a chronic course and the clinical manifestations are indistinguishable from those seen in cicatricial pemphigoid.

FIGURE 267

Linear IgA disease.  
Gingival lesions presenting  
as desquamative gingivitis



FIGURE 268

Linear IgA disease.  
Erythema and bullous formation in  
the form of desquamative gingivitis



FIGURE 269

Linear IgA disease.  
Small vesicles on the skin



- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Direct immunofluorescence.
  - ▶ Indirect immunofluorescence.

- Differential Diagnosis**
- ▶ Cicatricial pemphigoid
  - ▶ Bullous pemphigoid
  - ▶ Epidermolysis bullosa acquisita
  - ▶ Pemphigoid gestationis
  - ▶ Pemphigus

- Treatment**
- ▶ Local and systemic corticosteroids
  - ▶ Dapsone and sulphapyridine

### Pemphigoid Gestationis

- Definition**
- ▶ Pemphigoid gestationis or herpes gestationis is a distinct, rare and recurrent bullous dermatosis of pregnancy and the immediate postpartum period.

- Etiology**
- ▶ Unknown. It is currently thought to be immunologically mediated (autoimmune).

- Gingival Involvement**
- ▶ Very uncommon
  - ▶ Non-plaque-induced lesions with no attachment loss

- Other Sites of Involvement**
- ▶ Oral mucosa (buccal mucosa, tongue, palate), very rare
  - ▶ Skin (abdomen, chest, back, face, extremities, palms, soles)

- Main Clinical Features**
- ▶ Gingival lesions may appear as localized haemorrhagic bullae that very soon rupture leaving painful erosions (**Fig. 270**).
  - ▶ Multiple bullae that quickly rupture, leaving non-diagnostic painful erosions, may appear on the oral mucosa. The oral lesions always follow skin lesions and quickly retreat.

**FIGURE 270**

Pemphigoid gestationis.  
Haemorrhagic bullae on the gingiva





FIGURE 271

Pemphigoid gestationis.  
Numerous vesicles on the skin



- ▶ The skin manifestations are characterized by small papulovesicular lesions that are often grouped and commonly occur on a lesion of erythema and/or urticaria. Numerous widespread vesicles and crusting are common (Fig. 271). Characteristically, all these lesions are extremely pruritic.
- ▶ The eruptions usually appear during the second and the third trimester of pregnancy or immediately postpartum and resolve within 3–4 months after delivery.
- ▶ Rarely the disease may arise in association with trophoblastic malignancy.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Direct immunofluorescence.
  - ▶ Indirect immunofluorescence.

- Differential Diagnosis**
- ▶ Bullous pemphigoid
  - ▶ Linear IgA disease
  - ▶ Dermatitis herpetiformis
  - ▶ Pemphigus

- Treatment**
- ▶ Corticosteroid systematically. Azathioprine may also be used as adjuvant therapy.

### Epidermolysis Bullosa Acquisita

- Definition**
- ▶ Epidermolysis bullosa acquisita is a rare chronic mechanobullous mucocutaneous disease.

- Etiology**
- ▶ Autoimmune

- Gingival Involvement**
- ▶ Rare
  - ▶ Non-plaque-induced lesions with no attachment loss

FIGURE 272

Epidermolysis bullosa acquisita.  
Erythema and small vesicle  
on the gingiva in the form  
of desquamative gingivitis



FIGURE 273

Epidermolysis bullosa acquisita.  
Erythema and scarring on the skin



#### Other Sites of Involvement

- ▶ Oral mucosa (buccal mucosa, palate, tongue, lips).
- ▶ Skin, over the joints, extended.
- ▶ Other mucosae (conjunctival, nasal, oesophageal) are rarely involved.
- ▶ Surfaces of the knees, feet, elbows, ankles and hands, nails.

#### Main Clinical Features

- ▶ Gingival involvement may appear either in the form of desquamative gingivitis or as localized bullous formations that rupture, leaving quickly non-diagnostic painful erosions (**Fig. 272**).
- ▶ Localized and, rarely, widespread bullae and erosions may develop, particularly in friction-associated areas of the oral mucosa.
- ▶ The skin lesions are characterized by haemorrhagic bullae spontaneously or after mild trauma. The bullae rupture, leaving ulcers that may heal with scarring (**Fig. 273**). Milia, atrophic areas, hyperpigmentation and nail dystrophy may occur.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Direct immunofluorescence.
  - ▶ Electron microscopy.

- Differential Diagnosis**
- ▶ Cicatricial pemphigoid
  - ▶ Bullous pemphigoid
  - ▶ Linear IgA disease
  - ▶ Pemphigus

- Treatment**
- ▶ Local and systemic corticosteroid
  - ▶ Dapsone, azathioprine
  - ▶ Symptomatic and avoid trauma
  - ▶ Local and/or systemic antibiotics and antiseptics

### Dermatitis Herpetiformis

- Definition**
- ▶ Dermatitis herpetiformis, or Duhring-Brocq disease, is a rare chronic, recurrent pruritic vesicular skin disease.
  - ▶ The disease is more common between 20 and 50 years of age and males are more frequently affected than females.

- Etiology**
- ▶ Unknown. However, it is thought that immunological mechanisms may be involved in the pathogenesis.
  - ▶ A strong association with HLA-B<sub>8</sub>, HLAA<sub>1</sub>, HLA-DR<sub>3</sub>, HLA-DW<sub>3</sub>, HLA-DW<sub>2</sub> has been found.
  - ▶ Gluten hypersensitivity plays an important role in the pathogenesis.

- Gingival Involvement**
- ▶ Rare
  - ▶ Non-plaque-induced gingival lesions with no attachment loss

- Other Sites of Involvement**
- ▶ Oral mucosa (palate, tongue, buccal mucosa, lips) approximately 5%–10%
  - ▶ Skin (in a symmetrical distribution of the extensor surfaces of the elbows, knees, shoulders, buttocks, posterior neck, scale), always

- Main Clinical Features**
- ▶ Gingival and other oral mucosal lesions appear as localized whitish maculopapular lesions, erythema and formation of bullae that soon rupture, leaving non-specific painful erosions (**Figs. 274, 275**).
  - ▶ The oral lesions usually follow the skin eruption.
  - ▶ The skin lesions appear as erythematous papules or plaques accompanied by severe burning and pruritus as well as small vesicles coalescing in groups in a herpes-like pattern (**Fig. 276**).
  - ▶ The disease runs a very prolonged course with remissions and exacerbations.

- Diagnosis**
- ▶ The clinical diagnosis should always be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Direct immunofluorescence.
  - ▶ Indirect immunofluorescence.

- Differential Diagnosis**
- ▶ Bullous pemphigoid
  - ▶ Linear IgA disease
  - ▶ Pemphigoid gestations
  - ▶ Pemphigus



**FIGURE 274**

Dermatitis herpetiformis.  
Erythema and erosions  
on the gingiva and small bulla  
on the labial mucosa

**FIGURE 275**

Dermatitis herpetiformis.  
Whitish maculopapular lesions on  
the gingiva and the alveolar mucosa

**FIGURE 276**

Dermatitis herpetiformis.  
Group of vesicles on the skin  
in a herpes-like pattern



- ▶ Erythema multiforme
- ▶ Aphthous ulcers
- ▶ Herpetic gingivostomatitis

- Treatment**
- ▶ Gluten-free diet.
  - ▶ Sulphones and sulphapyridines.
  - ▶ Corticosteroid in persistent cases.
  - ▶ Tetracyclines and nicotinamide have also been used.

### Pemphigus Vulgaris

- Definition** ▶ Pemphigus vulgaris is a chronic mucocutaneous bullous disease.

- Etiology** ▶ Autoimmune

- Gingival Involvement**
- ▶ Common, approximately 20%–30%
  - ▶ Non-plaque-induced lesions with no attachment loss

- Other Sites of Involvement**
- ▶ Oral mucosa (soft palate, buccal mucosa, tongue, floor of the mouth, lips), very common 75%–90%
  - ▶ Other mucosae (conjunctiva, nose, genital, rectal, larynx, pharynx)
  - ▶ Skin

- Main Clinical Features**
- ▶ Gingival lesions may appear either in the form of desquamative gingivitis usually localized or as painful erosions on an erythematous base. Very rarely, gingival lesions may be the only site of involvement (Figs. 277–279).
  - ▶ The oral mucosa lesions present as erythema, bullous formation, painful erosions (Fig. 280).
  - ▶ The skin lesions present as bullous formation that soon rupture, leaving painful erosions.
  - ▶ Nikolsky's sign positive.

- Diagnosis**
- ▶ The clinical diagnosis should always be confirmed histologically and immunologically.
  - ▶ Biopsy and histopathological examination.
  - ▶ Direct immunofluorescence.
  - ▶ Indirect immunofluorescence.
  - ▶ Tzanck smear test.

- Differential Diagnosis**
- ▶ Cicatricial pemphigoid
  - ▶ Bullous pemphigoid
  - ▶ Pemphigoid gestations
  - ▶ Linear IgA disease
  - ▶ Dermatitis herpetiformis
  - ▶ Erythema multiforme
  - ▶ Epidermolysis bullosa acquisita
  - ▶ Herpetic gingivostomatitis
  - ▶ Aphthous ulcers

- Treatment**
- ▶ High dose of corticosteroids
  - ▶ Azathioprine, ciclosporin, mycophenolate mofetil as adjuvant treatment
  - ▶ Plasmapheresis

FIGURE 277

Pemphigus vulgaris.  
Early lesions on the gingiva  
presenting as erythema



FIGURE 278

Pemphigus vulgaris.  
Erythema and erosions on  
the gingiva in the form  
of desquamative gingivitis



FIGURE 279

Pemphigus vulgaris.  
Erosions on the gingiva and  
the alveolar mucosa





FIGURE 280

Pemphigus vulgaris.  
Extensive chronic erosions on the  
tongue and lower lip



### Lichen Planus

- Definition**
- ▶ Lichen planus is a common, chronic inflammatory mucocutaneous disease, affecting approximately 0.5%–2.5% of the population.
  - ▶ Females are affected more often than males, usually between 40 and 60 years of age.

- Etiology**
- ▶ Unknown. Current evidence suggests that there is a local cell-mediated immune response with a T lymphocyte infiltrate.

- Gingival Involvement**
- ▶ Common, approximately 25%–40%
  - ▶ Non-plaque-induced lesions with no attachment loss

- Other Sites of Involvement**
- ▶ Oral mucosa (buccal mucosa, tongue, lips, palate), approximately 50%–70% of the cases
  - ▶ Other mucosae (vulva, vagina, penis)
  - ▶ Skin (usually on the flexor surfaces of the forearms and wrists, the back, the sacral area, nails), commonly in a symmetrical pattern

- Main Clinical Features**
- ▶ Seven clinical forms of oral lichen planus have been described:
    - Papular.
    - Reticular.
    - Erosive.
    - Atrophic.
    - Hypertrophic.
    - Bullous.
    - Pigmented.
  - ▶ Most patients with oral lichen planus present with a combination of the various clinical forms. Polygonal white papules and lines are the basic clinical disorder of the disease.
  - ▶ Frequently the disease affects the gingiva and occasionally can be the initial or the only sign of oral involvement. Clinically, gingival lesions may appear in the form of desquamative gingivitis (erythema, oedema, desquamation of the epithelium or vesicle formation after pressure, and sensitive) (Figs. 281–283). White papules and lines, erosions or atrophy may also be found (Fig. 284).

**FIGURE 281**

Lichen planus.  
Erythema and white lines  
on the gingiva in the form of  
desquamative gingivitis

**FIGURE 282**

Lichen planus.  
Typical lesions of  
desquamative gingivitis

**FIGURE 283**

Lichen planus.  
Diffuse erythema on the  
palatal gingiva in the form of  
desquamative gingivitis





FIGURE 284

Lichen planus.  
Characteristic papules and lines  
on the gingiva



FIGURE 285

Lichen planus.  
Typical Wickham's striae  
on the buccal mucosa



- ▶ On the oral mucosa, small white papules that usually coalesce and form lines (Wickham's striae) and network of lines (**Fig. 285**). Annular distribution of the papules, painful erosions, atrophic or hypertrophic areas, bullous or pigmented lines may also be seen, depending on the clinical form.
- ▶ The skin lesions appear as small, red or violaceous flat, polygonal, shiny and pruritic papules.

#### Diagnosis

- ▶ The diagnosis is usually based on clinical criteria, but tests that may be supportive include:
- ▶ Biopsy and histopathological examination.
- ▶ Direct immunofluorescence.

#### Differential Diagnosis

- ▶ Cicatricial pemphigoid
- ▶ Lupus erythematosus
- ▶ Cinnamon contact stomatitis
- ▶ Lichen planus reaction due to drugs



- ▶ Lichenoid reaction to dental restorative material
- ▶ Graft-versus-host disease, lichen planus reactions
- ▶ Plasma cell gingivitis

- Treatment**
- ▶ Good oral hygiene and avoid pressure on the gingiva.
  - ▶ Corticosteroid topically in an adhesive base.
  - ▶ Corticosteroid systemically in severely painful lesions.
  - ▶ Dapsone, ciclosporin, tacrolimus and azathioprine may also be used in severe cases.

### Chronic Ulcerative Stomatitis

- Definition**
- ▶ Chronic ulcerative stomatitis is a rare autoimmune disease that resembles, clinically and histopathologically, lichen planus and lupus erythematosus but possesses a unique immunofluorescent pattern.

- Etiology**
- ▶ Autoimmune

- Gingival Involvement**
- ▶ Common
  - ▶ Non-plaque-induced gingival lesions with no attachment loss

- Other Sites of Involvement**
- ▶ Oral mucosa, commonly the buccal mucosa and the tongue

- Main Clinical Features**
- ▶ The gingival lesions usually appear in the form of desquamative gingivitis or as localized painful erythema and erosions (**Fig. 286**).
  - ▶ The oral mucosa lesions appear as painful erosions usually associated with white reticular lesions identical to those seen in oral lichen planus and discoid lupus erythematosus (**Fig. 287**).

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Direct immunofluorescence.
  - ▶ Indirect immunofluorescence.
  - ▶ Both in vivo binding to the oral mucosa and skin of a stratified epithelium-specific antinuclear antibody and high titres of these antibodies in serum are characteristic markers of this disease.

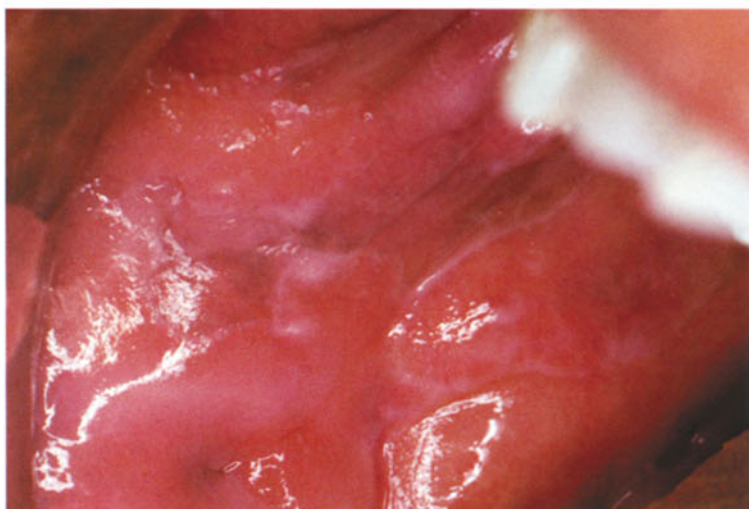
**FIGURE 286**

Chronic ulcerative stomatitis.  
Erythema and erosions  
on the gingiva in the form of  
desquamative gingivitis



FIGURE 287

Chronic ulcerative stomatitis.  
White lesions on the buccal mucosa  
mimic lichen planus

**Differential Diagnosis**

- ▶ Lichen planus
- ▶ Discoid lupus erythematosus
- ▶ Cicatricial pemphigoid
- ▶ Linear IgA disease
- ▶ Bullous pemphigoid
- ▶ Epidermolysis bullosa
- ▶ Pemphigus

**Treatment**

- ▶ Systemic or local corticosteroid
- ▶ Hydroxychloroquine sulphate (Plaquenil) and other antimalarial agents

**Psoriasis****Definition**

- ▶ Psoriasis is a common, chronic and recurring inflammatory skin disease.

**Etiology**

- ▶ Unknown

**Gingival Involvement**

- ▶ Very rare
- ▶ Non-plaque-induced lesions with no attachment loss

**Other Sites of Involvement**

- ▶ Oral mucosa (tongue, buccal mucosa, lips), rare (approximately 2%–4%)
- ▶ Skin (elbows, knees, scalp, lumbar area, nails)

**Main Clinical Features**

- ▶ The gingival lesions may resemble desquamative gingivitis; however, desquamation of the epithelium after pressure does not occur (Fig. 288). This pattern is an exception phenomenon.
- ▶ Oral lesions may appear as atypical erythema, white or greyish plaques and more often as circinate lesions mimicking geographic tongue. The oral lesions that usually develop after skin involvement are not diagnostic and a histopathological examination is needed to confirm diagnosis.

FIGURE 288

Psoriasis.  
Unusual involvement of the gingiva  
in the form of  
desquamative gingivitis



- ▶ Skin lesions are characterized by well-demarcated erythematous plaques with silvery scales on the surface. The lesions are usually asymptomatic.

**Diagnosis** ▶ The diagnosis is predominantly clinical but biopsy and histopathological examination are supportive.

**Differential Diagnosis** ▶ Geographic tongue and stomatitis  
▶ Cinnamon contact stomatitis  
▶ Lichen planus  
▶ Cicatricial pemphigoid

**Treatment** ▶ The gingival lesions should be treated with good oral hygiene and topical corticosteroids.  
▶ The treatment of the skin lesions must be left to the dermatologists.

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### Lupus Erythematosus

- Definition** ▶ Lupus erythematosus is a chronic inflammatory autoimmune disease with a variable spectrum of clinical forms, in which mucocutaneous clinical manifestations may occur in association with or without multiple organ systems involvement.
- Etiology** ▶ It is thought that a genetically susceptible autoimmune mechanism triggered by environmental factors is responsible for the pathogenesis of the disease.
- Classification** ▶ The disease is classified into two major forms:
- Discoid.
  - Systemic.
- Gingival Involvement** ▶ Rare
- ▶ Non-dental plaque-induced gingival lesions
- Other Involvement** ▶ Oral mucosa (15%–25% in discoid form and 30%–45% in systemic form)
- ▶ Other mucosae (conjunctiva, nose, genital)
- ▶ Skin
- ▶ Other organ systems (kidney, cardiovascular system, neurological system, joints)
- Main Clinical Features** ▶ The gingival lesions may appear either as localized erythema, usually associated with erosions that may occasionally mimic desquamative gingivitis (**Figs. 289, 290**). Less frequently lupus erythematosus may present as a non-specific painful ulceration surrounded by whitish spots or lines (**Fig. 291**).
- ▶ The oral mucosa lesions appear as well-defined atrophic red plaques usually surrounded by a sharp elevated border of radiating whitish striae. Erosions, atrophy, telangiectasias, white plaques and xerostomia may also occur.
- ▶ The skin lesions are characterized by flat or slightly elevated erythematous papules and plaques with scaling and follicular hyperkeratosis. Many patients in both forms develop the classic „butterfly“ rash on the face.
- ▶ Systemic lupus erythematosus is characterized by a broad spectrum of clinical and laboratory diagnostic criteria. Constitutional symptoms include fatigue, malaise, fever and weight loss associated with arthralgia and myalgia.



FIGURE 289

Lupus erythematosus.  
Irregular erythema and erosions as  
early lesions on the gingiva



FIGURE 290

Lupus erythematosus.  
Erosions on the gingiva



FIGURE 291

Lupus erythematosus.  
Localized erosions on the gingiva



- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination of oral and skin lesions.
  - ▶ Direct and indirect immunofluorescence.
- Differential Diagnosis**
- ▶ Lichen planus
  - ▶ Cicatricial pemphigoid
  - ▶ Bullous pemphigoid
  - ▶ Linear IgA disease
  - ▶ Pemphigus
  - ▶ Mixed connective tissue disease
- Treatment**
- ▶ Good dental plaque control.
  - ▶ Topical corticosteroids.
  - ▶ Antimalarials.
  - ▶ Systemic corticosteroids.
  - ▶ The treatment of systemic lupus erythematosus should be left to the specialist.

### Sjögren's Syndrome

- Definition**
- ▶ Sjögren's syndrome is a relatively common, chronic autoimmune disorder. The disease is more common in women between 40 and 60 years of age.
- Etiology**
- ▶ Obscure. However, genetic factors, viral infection and autoimmunity may be involved in the pathogenesis.
- Classification**
- ▶ Two forms are recognized:
    - Primary (sicca syndrome) with mainly oral and eye disorders.
    - Secondary, if it coexists with other autoimmune disease, usually rheumatoid arthritis.
- Gingival Involvement**
- ▶ Very rare
  - ▶ Dental plaque-induced gingival lesions modified by systemic factors
- Other Involvement**
- ▶ Salivary glands
  - ▶ Lacrimal glands
  - ▶ Other exocrine glands
  - ▶ Other organs and systems
- Main Clinical Features**
- ▶ Chronic gingivitis, localized or generalized due to xerostomia and/or candida infection may occur (**Fig. 292**). Rarely increased alveolar bone loss may also be seen.
  - ▶ Xerostomia is the hallmark of the disease. The oral mucosa is reddish, dry, smooth, and shiny (**Fig. 293**). Oral soreness, discomfort and dysphagia are common symptoms. Candidiasis, angular cheilitis, and dental caries are relatively common. Recurrent swelling of parotid and other major salivary glands may occur.
  - ▶ The ocular manifestations characterized by dryness of the eyes, keratoconjunctivitis sicca and more severe lesions as the disease progress
  - ▶ In the secondary form of the disease, the clinical manifestations depend on the associated autoimmune diseases.

FIGURE 292

Sjögren's syndrome.  
Generalized chronic gingivitis



FIGURE 293

Sjögren's syndrome.  
Dry, reddish and shiny dorsum  
of the tongue

**Diagnosis**

- ▶ The clinical diagnosis should be confirmed by laboratory tests.
- ▶ Biopsy and histopathological examination of the minor salivary glands.
- ▶ Serological tests:
  - Antinuclear antibodies.
  - Anti-SSA (Ro).
  - Anti-SSB (La) antibodies.
  - Rheumatoid factor.

**Differential Diagnosis**

- ▶ Graft-versus-host disease
- ▶ Xerostomia due to drugs
- ▶ Systemic sclerosis
- ▶ Mixed connective tissue disease

**Treatment**

- ▶ Good dental plaque control
- ▶ Saliva and tear substitutes
- ▶ Antimalarial drugs, corticosteroid, immunosuppressants



## Systemic Sclerosis

- Definition** ▶ Systemic sclerosis is a chronic multisystemic inflammatory connective tissue disorder with a broad spectrum of manifestations.
- Etiology** ▶ Obscure. However, an autoimmune pathogenesis may be involved. In vitro studies suggest that the products of the autoimmune reaction may influence the behaviour of fibroblasts and endothelial cells leading to the overproduction of connective tissue by fibroblasts.
- Gingival Involvement** ▶ Rare  
▶ Non-dental plaque-induced gingival lesions
- Other Involvement** ▶ Oral mucosa (relatively common)  
▶ Skin (always)  
▶ Viscera (esophagus, intestinal tract, kidney, lungs, heart, skeletal muscles)
- Main Clinical Features** ▶ The gingival lesions are minor and characterized by pallor of free and attached gingiva, gingival regression and very rarely greater evidence of periodontitis (**Fig. 294**). However, other studies revealed no significant differences in periodontal status between systemic sclerosis patients and healthy controls.
- ▶ The oral mucosal lesions include:
- Xerostomia.
  - Thin and pale oral mucosa.
  - Short and hard lingual frenulum.
  - Atrophy of the tongue papillae.
  - Microglossia.
  - Microstomia.
  - Limited mouth opening.
  - Perioral radial folds.
- ▶ On dental radiographs, a diffuse widening of the periodontal ligament space may be seen approximately in 10%–20% of patients. Difficulty in swallowing, dysphagia are also common.
- ▶ The skin lesions are characterized initially by oedema but as the disease progresses the skin becomes thin, hard, inelastic and pale. Facial

**FIGURE 294**

Systemic sclerosis.  
Pallor and thin gingivae



skin involvement results in the characteristic mask-like facies. Skin ulcers, telangiectasias, Raynaud's phenomenon are also common.

- ▶ CREST syndrome (Calcinosis cutis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, telangiectasia) is a clinical limited variant of systemic sclerosis.
- ▶ Visceral involvement usually leads to organ failure.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Skin biopsy and histopathological examination.
  - ▶ Immunological examinations.

- Differential Diagnosis**
- ▶ Epidermolysis bullosa
  - ▶ Cicatricial pemphigoid
  - ▶ Lipoid proteinosis

- Treatment**
- ▶ Good oral hygiene
  - ▶ Systemic treatment includes:
    - Antimalarials
    - Corticosteroids
    - Immunosuppressive drugs
    - D-penicillamine
    - Colchicine
    - Others

### Mixed Connective Tissue Disease

- Definition**
- ▶ Mixed connective tissue disease is a multisystemic disorder characterized by the simultaneous clinical manifestations mainly seen in lupus erythematosus, systemic sclerosis and polymyositis.

- Etiology**
- ▶ The cause and pathogenesis remain obscure.

- Gingival Involvement**
- ▶ Rare
  - ▶ Non-dental plaque-induced gingival lesions

- Other Involvement**
- ▶ Oral mucosa, rare
  - ▶ Skin
  - ▶ Skeletal muscles, lymph nodes and several other organs

- Main Clinical Features**
- ▶ The gingival lesions may appear as localized or diffuse erythema and/or non-specific irregular erosions identical to those seen in lupus erythematosus patients (Fig. 295). Periodontal destruction is impossible.
  - ▶ The oral mucosal lesions are characterized by a non-specific erythema, oedema, erosions and xerostomia.
  - ▶ The skin and other manifestations are those seen in systemic lupus erythematosus, systemic scleroderma and polymyositis.

- Diagnosis**
- ▶ The clinical diagnosis should always be confirmed by laboratory tests.
  - ▶ High titres of anti-ribonucleoprotein antibodies (anti-RNP) is the most diagnostic laboratory finding.
  - ▶ Other serological tests specific for the group of diseases included.

- Differential Diagnosis**
- ▶ Lupus erythematosus
  - ▶ Systemic scleroderma

FIGURE 295

Mixed connective tissue disease.  
Irregular erosions on the gingiva



- ▶ Polymyositis
- ▶ Other connective tissue diseases

#### Treatment

- ▶ Good oral hygiene
- ▶ Topical corticosteroids
- ▶ Systemic treatment includes:
  - Corticosteroids
  - Immunosuppressive drugs
  - Antimalarials

### Graft-Versus-Host Disease

#### Definition

- ▶ Graft-versus-host disease (GVHD) is a multisystemic phenomenon due to immune reaction between the transplanted T lymphocytes and the patient's normal tissues.

#### Etiology

- ▶ The disease occurs in recipients of allogeneic bone marrow transplants usually to treat life-threatening diseases of the blood or bone marrow.

#### Classification

- ▶ Two forms of the disorder are recognized:
  - Acute, that is typically observed within 100 days of transplantation
  - Chronic, that develops later than 100 days after transplantation

#### Gingival Involvement

- ▶ Rare
- ▶ Non-dental plaque-induced gingival lesions

#### Other Involvement

- ▶ Oral mucosa (40 %–80 %)
- ▶ Skin, common
- ▶ Gastrointestinal tract, liver, lungs, eyes

#### Main Clinical Features

- ▶ The gingival lesions are not specific and are characterized by diffuse or localized erythema usually associated with a fine, reticular network of white striae and/or erosions similar to those seen in lichen planus (**Fig. 296**). The condition occasionally mimics desquamative gingivitis.



FIGURE 296

Graft-versus-host disease.  
Diffuse erythema on the gingiva  
mimic desquamative gingivitis



FIGURE 297

Graft-versus-host disease.  
Lichenoid lesions on the  
buccal mucosa



- ▶ The oral mucosal lesions appear as diffuse erythema, lichenoid pattern, and painful ulcerations (**Fig. 297**). Atrophy of the oral mucosa, a burning sensation and xerostomia are common. The oral mucosal and gingival lesions are more common in the chronic form of the disorder.
- ▶ The skin lesions range from mild rash with hyperpigmentation to lesions that may resemble to systemic sclerosis and lichen planus.
- ▶ Other signs and symptoms are nausea, vomiting, abdominal pain, diarrhea, liver dysfunction.

#### Diagnosis

- ▶ The diagnosis is usually based on the clinical criteria and less on laboratory tests.
- ▶ Biopsy and histopathological examination (oral mucosa and the minor salivary glands, skin).
- ▶ Increased salivary sodium concentration.

#### Differential Diagnosis

- ▶ Lichen planus
- ▶ Lupus erythematosus

- ▶ Systemic sclerosis
- ▶ Sjögren's syndrome
- ▶ Drug reactions
- ▶ Leukaemia

- Treatment**
- ▶ Good dental plaque control
  - ▶ Topical corticosteroids
  - ▶ Systemic corticosteroids, immunosuppressive drugs, thalidomide

### Behçet's Disease

- Definition**
- ▶ Behçet's disease is a chronic multisystemic disorder that affects more commonly certain ethnic groups (Japanese, Turkish, Greek and other Mediterranean origins).

- Etiology**
- ▶ The exact etiology remains obscure. However, genetic, immunological factors, and viral infection may be involved in the pathogenesis of the disease.

- Gingival Involvement**
- ▶ Very rare
  - ▶ Non-dental plaque-induced gingival lesions

- Other Involvement**
- ▶ Oral mucosa, very common
  - ▶ Other mucosae (eyes, genitalia, intestinal)
  - ▶ Skin
  - ▶ Joints, cardiovascular systems, lungs, central nervous system

- Main Clinical Features**
- ▶ The main oral manifestations of Behçet's disease are recurrent aphthous ulcerations (minor, major, herpetiform, or atypical). Painful (1–2 in number) aphthous ulcers, usually minor or atypical, may rarely develop, usually on the attached gingiva (**Figs. 298, 299**).
  - ▶ Conjunctivitis, iritis, iridocyclitis with hypopyon, uveitis are the most frequent eye manifestations.
  - ▶ The skin lesions present as genital round ulcerations on the scrotum or on the penis or vulva (**Fig. 300**). Folliculitis, papules, pustules, nodular erythema and less often necrotic lesions are common cutaneous lesions.

**FIGURE 298**

Behçet's disease.  
Aphthous ulcers on the upper gingiva



FIGURE 299

Behçet's disease.  
Aphthous ulcers on the upper gingiva



FIGURE 300

Behçet's disease.  
Typical round ulcers on the scrotum.



- ▶ Vascular, intestinal, joint and neurological involvement may occur.
- ▶ Pathergy test is positive in about 60%–70% of cases.

**Diagnosis**

- ▶ The diagnosis is made on clinical criteria.
- ▶ Biopsy and histopathological examinations may be supportive.

**Differential Diagnosis**

- ▶ Aphthous ulcers
- ▶ Pemphigus
- ▶ Pemphigoid
- ▶ Erythema multiforme
- ▶ Necrotizing ulcerative gingivitis
- ▶ Neutropenia
- ▶ Leukaemia
- ▶ Langerhans cell histiocytosis

**Treatment**

- ▶ Corticosteroids topically
- ▶ Systemic corticosteroids



- ▶ Immunosuppressive drugs
- ▶ Thalidomide
- ▶ Colchicine

### Reiter's Disease

- Definition** ▶ Reiter's disease is an uncommon multisystemic disorder that may be triggered by an infectious agent in a genetically susceptible individual.
- Etiology** ▶ The exact etiology remains unknown, although the pathogenesis is mediated by an immunological mechanism.
- Gingival Involvement** ▶ Rare  
▶ Non-dental plaque-induced gingival lesions
- Other Involvement** ▶ Oral mucosa (20%–40%)  
▶ Genital, eyes, skin, joints, skeletal, cardiovascular system
- Main Clinical Features** ▶ The gingival lesions appear as localized or generalized non-specific erythema, usually on the attached gingiva, occasionally associated with superficial, slightly sensitive erosions (Figs. 301, 302).  
▶ The oral mucosal lesions are characterized by diffuse erythematous areas intermixed with thin whitish dots or lines, and superficial erosions. Localized loss of the tongue filiform papillae in the form of geographic tongue may occur (Fig. 303)  
▶ The skin manifestations appear as macular, vesicular or pustular, psoriasiform lesions and keratoderma.  
▶ Other manifestations include conjunctivitis, cyclic balanitis, non-gonococcal urethritis, prostatitis, cervicitis and nail disturbances.  
▶ The most characteristic feature of the disease is the symmetrical arthritis of six to seven joints.
- Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.  
▶ Skin biopsy and histopathological examination.  
▶ Blood and serological tests, HLA antigens, synovial fluid analysis.

FIGURE 301

Reiter's disease.  
Diffuse deep erythema and erosions  
on the upper gingiva



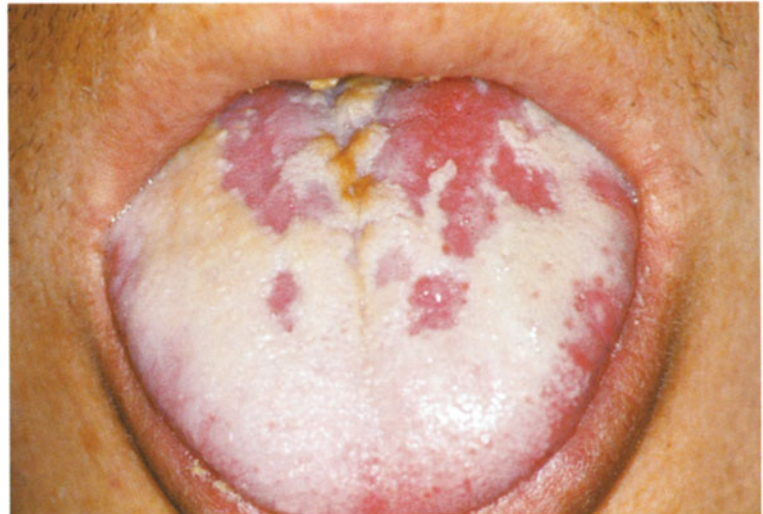
FIGURE 302

Reiter's disease.  
Slight erythema and oedema  
on the upper gingiva



FIGURE 303

Reiter's disease.  
Localized loss of the filiform papillae  
of the dorsum of the tongue

**Differential Diagnosis**

- ▶ Behçet's disease
- ▶ Erythema multiforme
- ▶ Geographic stomatitis
- ▶ Drug eruption

**Treatment**

- ▶ Non-steroidal anti-inflammatory drugs, salicylates
- ▶ Corticosteroids

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## Orofacial Granulomatosis

### Melkersson-Rosenthal Syndrome

- Definition** ▶ Melkersson-Rosenthal syndrome is a rare disorder. It consists of the triad of recurrent facial swelling, facial paralysis and fissured tongue.
- Etiology** ▶ Unknown
- Gingival Involvement** ▶ Rare  
▶ Non-dental plaque-induced gingival lesions
- Other Involvement** ▶ Oral mucosa  
▶ Facial
- Main Clinical Features** ▶ Gingival lesions present as irregular, oedematous swelling, slightly erythematous, affecting mainly the anterior interdental papillae and free and attached gingiva (**Figs. 304–306**).
- ▶ Granulomatous cheilitis presenting as diffuse recurrent swelling of the lips is the cardinal feature of the syndrome. Swelling of the lips is usually the first sign. After a long period of attacks, the swelling persists and may become permanent.
- ▶ Buccal, palatal and lingual swelling may also occur (**Fig. 307**).
- ▶ Facial nerve palsy, similar to Bell's palsy is present in 20%–30% of the cases and is frequently associated with facial oedema. Facial nerve palsy may be the first sign and is usually unilateral.
- ▶ Fissured tongue is present in about half of the patients.
- ▶ The disease affects equally both sexes and usually young individuals.
- Diagnosis** ▶ The diagnosis is based on clinical and laboratory criteria.  
▶ Allergy tests.  
▶ Biopsy and histopathological examination of oral lesions.
- Differential Diagnosis** ▶ Angioedema  
▶ Cheilitis glandularis  
▶ Crohn's disease  
▶ Sarcoidosis  
▶ Amyloidosis
- Treatment** ▶ Dental plaque control and surgical excision of the gingival lesions.  
▶ Corticosteroid, topical or systemic.  
▶ If facial nerve palsy is present, neurological consultation should be undertaken.

FIGURE 304

Melkersson-Rosenthal syndrome.  
Early gingival lesions mimic  
chronic gingivitis



FIGURE 305

Melkersson-Rosenthal syndrome.  
Localized gingival swelling



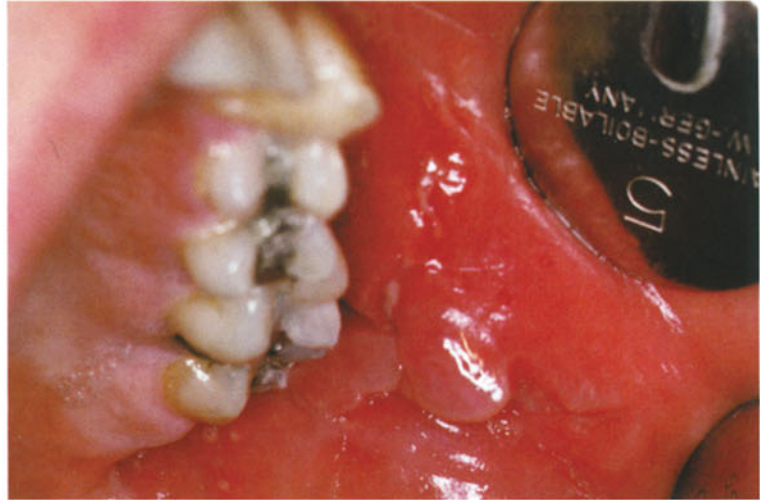
FIGURE 306

Melkersson-Rosenthal syndrome.  
Diffuse gingival and labial swelling



FIGURE 307

Melkersson-Rosenthal syndrome.  
Multiple oedematous folds  
on the buccal mucosa



### Crohn's Disease

**Definition** ▶ Crohn's disease is a chronic granulomatous inflammatory disease that may affect any part of the gastrointestinal tract from the oral cavity to the anus.

**Etiology** ▶ Unknown. A hereditary susceptibility is possible and an immunologically mediated mechanism may be involved in the pathogenesis.

**Gingival Involvement** ▶ Relative common  
▶ Non-dental plaque-induced gingival lesions

**Other Involvements** ▶ Oral mucosa, common  
▶ Gastrointestinal tract, particularly the terminal ileum, always  
▶ Skin, eyes, joints (rare)

**Main Clinical Features** ▶ The gingival lesions present as erythematous and oedematous gingival enlargement, which may be localized or generalized, usually affecting the anterior gingiva (**Figs. 308–310**). Rarely gingival involvement may be the presenting feature.  
▶ The oral mucosal lesions may manifest as diffuse granulomatous lip swelling, nodular or diffuse swelling of the tongue, soft palate and buccal mucosa, resulting in a cobblestone pattern. Mucosal folds and tags, lip fissures, aphthous or aphthous-like ulcerations, soft tissue granulomas, metallic dysgeusia, and persistent lymph node enlargement may occur.  
▶ The gastrointestinal signs and symptoms include abdominal discomfort, diarrhoea, vomiting, anorexia, weight loss, low-grade fever and rectal bleeding. These signs and symptoms may persist for many years with periods of exacerbation and remission.  
▶ Extra-abdominal manifestations include arthritis, spondylitis and uveitis.

**Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.  
▶ Biopsy and histopathological examination of oral and gut lesions.



FIGURE 308

Crohn's disease.  
Localized gingival swelling



FIGURE 309

Crohn's disease.  
Generalized swelling  
on the upper gingiva



FIGURE 310

Crohn's disease.  
Oedematous swelling of the  
lower gingiva and lips



- Differential Diagnosis**
- ▶ Melkersson-Rosenthal syndrome
  - ▶ Sarcoidosis
  - ▶ Ulcerative colitis
  - ▶ Pyostomatitis vegetans
  - ▶ Contact hypersensitivity reactions
- Treatment**
- ▶ Good oral hygiene
  - ▶ Topical corticosteroid for oral lesions
  - ▶ Systemic corticosteroids, sulphasalazine, azathioprine, ciclosporin, metronidazole for gut lesions

### Sarcoidosis

- Definition**
- ▶ Sarcoidosis is a chronic multisystemic granulomatous disease affecting one or multiple organs or tissues.
- Etiology**
- ▶ Unknown. However, the disease is usually associated with depression of cell-mediated immunity and an overactivity of B cells.
- Gingival Involvement**
- ▶ Rare
  - ▶ Non-dental plaque-induced gingival lesions
- Other Involvement**
- ▶ Oral mucosa, rare
  - ▶ Lymph nodes, salivary glands, skin, eyes, bone, nerves
  - ▶ Lungs, liver, spleen
- Main Clinical Features**
- ▶ The gingival lesions usually present as diffuse erythema extending to alveolar mucosa (**Fig. 311**). The affected gingiva is slightly oedematous and sensitive, and occasionally may be the initial sign of sarcoidosis (**Fig. 312**).
  - ▶ The oral lesions present as painless, solitary or multiple, red nodules that may rarely ulcerate. Lip and less often buccal and tongue swelling may occur. Salivary gland swelling (minor and major), temporomandibular dysfunction, facial nerve palsy may also occur.
  - ▶ The skin lesions include painless, purple-brown macules, papules or nodules that may be scattered or confluent, lupus pernio, erythema nodosum, scars and plaques. The skin manifestations appear in 25%–30% of cases and may remain the only lesions for long time.
  - ▶ Lymphadenopathy, liver and spleen enlargement, lung disturbances are common.
- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Serum levels of angiotensin-converting enzyme (SACE).
  - ▶ Chest radiography.
  - ▶ Gallium scan.
- Differential Diagnosis**
- ▶ Crohn's disease
  - ▶ Tuberculosis
  - ▶ Amyloidosis
  - ▶ Melkersson-Rosenthal syndrome
- Treatment**
- ▶ Good oral hygiene and topical corticosteroid for gingival lesions
  - ▶ Systemic corticosteroids, allopurinol for systemic manifestations

FIGURE 311

Sarcoidosis.  
Diffuse erythema and  
slight oedema on the gingiva



FIGURE 312

Sarcoidosis.  
Severe oedema and swelling  
of the upper gingiva



### Pyostomatitis Vegetans

**Definition** ▶ Pyostomatitis vegetans is a relatively rare, pustular disorder of the mouth associated with inflammatory bowel diseases, particularly ulcerative colitis and Crohn's disease.

**Etiology** ▶ Unknown

**Gingival Involvement** ▶ Common (labial and buccal gingiva)  
▶ Non-dental plaque-induced gingival lesions

**Other Involvement** ▶ Oral mucosa (buccal, labial, mucolabial and mucobuccal folds, soft palate, ventral tongue)  
▶ Skin (pyodermitis vegetans is the equivalent of oral lesions)



FIGURE 313

Pyostomatitis vegetans.  
Early lesions on the gingiva  
presenting with erythema  
and oedema



FIGURE 314

Pyostomatitis vegetans.  
Erythema and multiple pustular  
microabscesses on the  
upper gingiva

**Main Clinical Features**

- ▶ The gingival lesions present as multiple, painless, yellow-white, vegetative mucosal folds that consist of pustular microabscesses that finally may be ulcerated (Figs. 313, 314).
- ▶ Similar lesions may be present in other mucosal sites.
- ▶ Rarely similar lesions may be present on the skin.

**Diagnosis**

- ▶ The clinical diagnosis should be confirmed by laboratory tests.
- ▶ Biopsy and histopathological examination.
- ▶ Direct immunofluorescence to rule out chronic bullous oral diseases.

**Differential Diagnosis**

- ▶ Pemphigus vegetans
- ▶ Dermatitis herpetiformis
- ▶ Wegener's granulomatosis

**Treatment**

- ▶ Systemic corticosteroids, sulphasalazine.
- ▶ Spontaneous remission of oral lesions may occur when bowel lesions subside.

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### Porphyria

- Definition** ▶ Porphyria comprises a group of disorders resulting from abnormal porphyrin metabolism.
- Etiology** ▶ Inborn enzymatic defects in the halm biosynthetic pathway. It is inherited as an autosomal dominant and recessive trait.
- Classification** ▶ Three major groups, each with several types are recognized:
- Erythropoietic porphyria.
  - Hepatic porphyria .
  - Erythropoietic-hepatic porphyria.
- ▶ Gingival lesions occurs only in erythropoietic protoporphyria and porphyria cutanea tarda.
- Gingival Involvement** ▶ Uncommon
- ▶ Non-dental plaque-induced gingival lesions
- Other Involvement** ▶ Oral mucosa, rare
- ▶ Skin, common
- ▶ Internal organ
- Main Clinical Features** ▶ The gingival lesions present as diffuse erythema of free and attached vestibular gingiva, occasionally mimicking desquamative gingivitis (**Fig. 315**).

**FIGURE 315**

Porphyria cutanea tarda.  
Diffuse erythema  
on the lower gingiva mimics  
desquamative gingivitis





- ▶ The oral mucosal lesions present as diffuse erythema, atrophy, vesicles and ulcerations, especially on the lips.
- ▶ The skin lesions appear mainly on the light-exposed areas and are characterized by photosensitivity, erythema, fragility, vesicles, ulcerations and scar formation, hypertrichosis.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination of oral and skin lesions.
  - ▶ Biochemical tests (erythrocyte, urine, faecal).
  - ▶ Photobiological tests and direct immunofluorescence.

- Differential Diagnosis**
- ▶ Desquamative gingivitis due to epidermolysis bullosa and cicatricial pemphigoid
  - ▶ Pellagra

- Treatment**
- ▶ Dental plaque control.
  - ▶ Systemic therapy must be left to the specialist.

### Amyloidosis

- Definition**
- ▶ Amyloidosis is a relatively rare metabolic disorder characterized by the extracellular deposition of an amorphous proteinaceous substance known as amyloid.

- Etiology**
- ▶ Unknown

- Classification**
- ▶ Four major forms are recognized:
    - Primary.
    - Secondary.
    - Familial.
    - Senile.
  - ▶ Oral lesions present mainly in primary systemic form.

- Gingival Involvement**
- ▶ Relatively rare
  - ▶ Non-dental plaque-induced gingival lesions

- Other Involvement**
- ▶ Oral mucosa (tongue, lips, buccal), common, particularly in primary form
  - ▶ Skin
  - ▶ Gastrointestinal tract, joints, skeletal muscles
  - ▶ Heart, kidneys and rarely other organs

- Main Clinical Features**
- ▶ The gingival lesions usually present as localized painless nodules or diffuse swelling with characteristic reddish colour (**Figs. 316, 317**).
  - ▶ The most common oral mucosal manifestations are petechiae, ecchymoses, nodules, haemorrhagic bullae, ulcers, macroglossia, minor and major salivary gland enlargements, xerostomia, and regional lymph node swelling (**Fig. 318**). The oral lesions may be the early presenting signs of the disease.
  - ▶ The most common skin lesions are purpura, petechiae, papules, nodules, bullae, ulcers, and alopecia.
  - ▶ The most common presenting symptoms are weakness, fatigue, weight loss, oedema, dyspnea, hoarseness, bleeding, pain, carpal tunnel syndrome.
  - ▶ About 20%–30% of the cases of primary amyloidosis are associated with multiple myeloma. The prognosis is unfavourable.

FIGURE 316

Amyloidosis.  
Localized red swelling on the gingiva



FIGURE 317

Amyloidosis.  
Gingival swelling on the  
upper gingiva



FIGURE 318

Amyloidosis.  
Macroglossia in a patient with  
primary systemic amyloidosis





- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination with special stains, e.g. Congo red, Sirius red, methyl violet, thioflavine-T.
  - ▶ Serum and urine electrophoresis.

- Differential Diagnosis**
- ▶ Soft tissue, gingival plasmacytoma
  - ▶ Sarcoidosis and Crohn's disease
  - ▶ Benign and malignant neoplasms of the gingiva, e.g. Kaposi's sarcoma, peripheral giant cell granuloma

- Treatment**
- ▶ Symptomatic treatment of gingival and oral lesions.
  - ▶ Systemic treatment must be left to the specialist.

### Langerhans Cell Histiocytosis

- Definition**
- ▶ Langerhans cell histiocytosis is a clinicopathological heterogeneous disorder characterized by proliferation of Langerhans cells.

- Etiology**
- ▶ The cause remains obscure.

- Classification**
- ▶ Three major forms are recognized:
    - Eosinophilic granuloma, that is a localized benign form of the disorder, usually characterized by solitary or multiple bone lesions and soft tissue lesions as well.
    - Hand-Schüller-Christian disease, that is a chronic disseminated form involving primarily bone, soft tissue and viscera.
    - Letterer-Siwe disease, that is an acute disseminated form involving skin, viscera, bone marrow.

- Periodontal Involvement**
- ▶ Relatively common, particularly in eosinophilic granuloma and Hand-Schüller-Christian disease
  - ▶ Periodontitis associated with systemic diseases

- Other Involvement**
- ▶ Oral mucosa, jaws
  - ▶ Bones, bone marrow, skin, visceral

- Main Clinical Features**
- ▶ The gingival lesions present as localized or multiple atypical ulcerations, usually associated with bone destruction and teeth loosening (Figs. 319–322). The gingival lesions may be the only manifestation for a long time, particularly in eosinophilic granuloma.
  - ▶ The oral mucosa lesions are less common and are characterized by atypical deep ulcerations, ecchymoses, oedema, halitosis, taste disturbances and are more common in Hand-Schüller-Christian and Letterer-Siwe forms.
  - ▶ Jaw bone destruction, either solitary or multiple, is common, in all three forms of the disorder.
  - ▶ Other bone involvement, exophthalmos, diabetes insipidus, skin rash, otitis media, hepatomegaly, splenomegaly, lymphadenopathy are common in the two severe forms of the disorder.
  - ▶ More than 50 % of all cases of the Langerhans cell histiocytosis are seen in patients under 10 years of age.

- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ Biopsy and histopathological examination.
  - ▶ Immunohistochemical examinations.
  - ▶ Radiographic examination.



**FIGURE 319**

Langerhans cell histiocytosis.  
Localized swelling on the  
upper gingiva in a patient with  
eosinophilic granuloma

**FIGURE 320**

Langerhans cell histiocytosis.  
Localized ulceration on the  
lower gingiva in a patient with  
eosinophilic granuloma

**FIGURE 321**

Langerhans cell histiocytosis.  
Localized gingival ulceration  
and bone destruction in a patient  
with eosinophilic granuloma

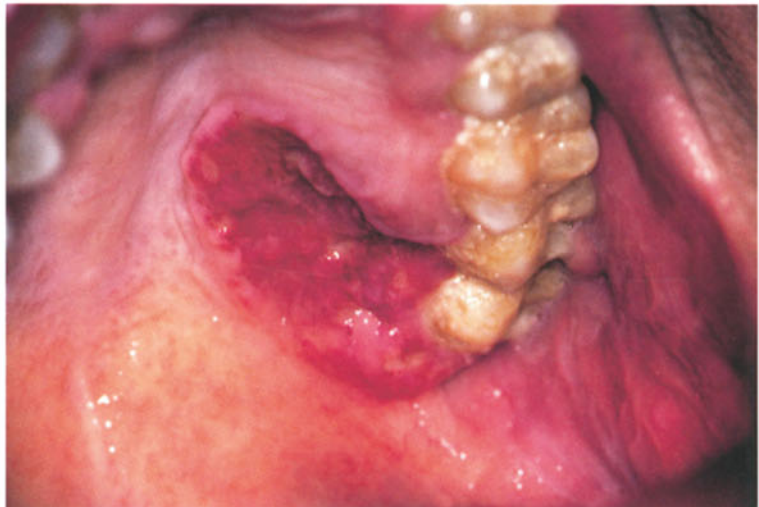
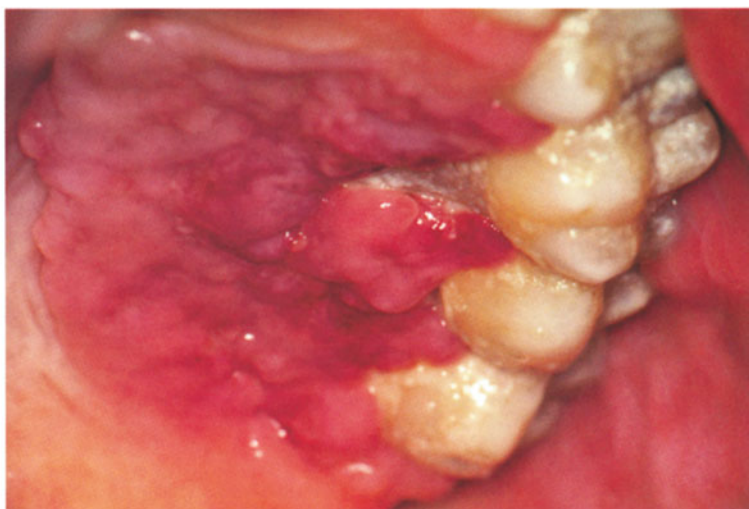


FIGURE 322

Langerhans cell histiocytosis.  
Extensive gingival and palatal  
ulcerations and bone  
destruction in a patient with  
Hand-Schüller-Christian disease



#### Differential Diagnosis

- ▶ Necrotizing ulcerative gingivitis and periodontitis
- ▶ Aggressive periodontitis
- ▶ Traumatic lesions on the gingiva
- ▶ Hypophosphatasia, acatalasia
- ▶ Haematological disorders
- ▶ Eosinophilic ulcer

#### Treatment

- ▶ Dental plaque control and periodontal curettage
- ▶ Radiation of local bone lesions
- ▶ Corticosteroid and chemotherapy for the systemic manifestations, which should be left to the specialists

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### Diabetes Mellitus

- Definition** ▶ Diabetes mellitus is a chronic, systemic, heterogeneous group of disorders of the metabolism of carbohydrate, protein and fat, resulting in microvascular and macrovascular complications.
- Etiology** ▶ It is due either to decreased production of insulin, tissue resistance to the effect of insulin, or a combination of both.
- Classification** ▶ Two main forms of diabetes mellitus are recognized:
- Type 1, insulin dependent (children onset).
  - Type 2, non-insulin dependent (adult onset).
- Gingival Involvement** ▶ Common, particularly in type 1
- ▶ Dental plaque-induced gingivitis and periodontitis, modified by diabetes mellitus
- Other Involvement**
- ▶ Oral mucosa, relatively rare
  - ▶ Many organs and systems
- Main Clinical Features**
- ▶ The clinical features of gingivitis associated with diabetes mellitus are characterized by the presence of dental plaque at gingival margin, erythema, oedema, bleeding upon probing, increased gingival exudate and the absence of attachment and alveolar bone loss (Figs. 323, 324). The gingival inflammation is usually reversible after the control of diabetic state. In type 2 diabetes mellitus patients, periodontitis with attachment loss and alveolar bone loss may develop.
  - ▶ The oral mucosa manifestations are not specific and include xerostomia, superficial erosions, retardation of wound healing, burning mouth, taste disturbances, candidiasis.
  - ▶ Salivary gland swelling (sialosis).
- Diagnosis** ▶ The clinical diagnosis should be confirmed by laboratory tests.
- ▶ Blood examination for sugar.
- Differential Diagnosis** ▶ Dental plaque-induced gingivitis
- ▶ Sex hormone-associated gingivitis
- Treatment** ▶ Dental plaque control
- ▶ Diabetic control from the specialist



FIGURE 323

Diabetes mellitus.  
Early periodontitis



FIGURE 324

Diabetes mellitus.  
Severe periodontitis



### Sex Steroid Hormones

**Definition** ▶ Sex steroid hormones (androgens, estrogens and progestins) may influence the periodontal tissue, and particularly the gingiva, to increase the inflammation response present.

**Etiology** ▶ Dental plaque gingivitis modified by sex steroid hormones

**Classification** ▶ Four forms of gingival disorders may be developed:

- Puberty-associated gingivitis.
- Menstrual cycle-associated gingivitis.
- Pregnancy-associated gingivitis.
- Oral contraceptive-associated gingivitis.

**Gingival Involvement** ▶ Common  
▶ Dental plaque-induced modified by sex steroid hormones

**FIGURE 325**

Early pregnancy-associated  
gingivitis

**FIGURE 326**

Severe pregnancy-associated  
gingivitis and granulomas

**FIGURE 327**

Severe pregnancy-associated  
gingivitis and granulomas



- Main Clinical Features**
- ▶ All four forms of gingivitis display a similar clinical pattern.
  - ▶ The main clinical features are dental plaque present at gingival margin, redness and oedema of the gingiva (**Figs. 325–327**), bleeding upon probing, loss of attachment and alveolar bone loss. The lesions can resolve following puberty, ovulation, at parturition, and discontinuation of oral contraceptives.
- Diagnosis**
- ▶ The diagnosis is based on the clinical criteria and laboratory tests.
  - ▶ Sex hormone measurement.
- Differential Diagnosis**
- ▶ Dental plaque-induced gingivitis
  - ▶ Diabetes mellitus-associated gingivitis
- Treatment**
- ▶ Dental plaque control
  - ▶ Balance of sex steroid hormones by the specialist

### Addison's Diseases

- Definition**
- ▶ Addison's disease is a rare acquired deficiency in adrenal gland function characterized by insufficient secretion of glucocorticoids and mineralocorticoids.
- Etiology**
- ▶ Destruction of the adrenal cortex due to infection, autoimmunity, infiltration by a tumour, haemorrhage, or infarction. The levels of ACTH and melanocyte-stimulating hormone (MSH) are elevated, causing increased hyperpigmentation of the skin and oral mucosa.
- Gingival Involvement**
- ▶ Relatively common
  - ▶ Non-dental plaque-induced gingival lesions
- Other Involvement**
- ▶ Oral mucosa (buccal, palate, lips), common
  - ▶ Skin, common
- Main Clinical Features**
- ▶ The gingival lesions present as multiple melanotic macules or irregular plaques (**Figs. 328, 329**).
  - ▶ Similar lesions appear on the other areas of the oral mucosa. The oral manifestations are usually early and valuable diagnostic sign.
  - ▶ The skin lesions present as diffuse bronze to grey hyperpigmentation, particularly on sun-exposed skin.
  - ▶ Malaise, weakness, nausea, vomiting and weight loss are common.
- Diagnosis**
- ▶ The clinical diagnosis should be confirmed by laboratory tests.
  - ▶ ACTH measurement, ACTH stimulation, and assay of plasma cortisol.
- Differential Diagnosis**
- ▶ Normal gingival melanosis
  - ▶ Smoking melanosis
  - ▶ Pigmentation due to drugs
  - ▶ Amalgam tattoo
  - ▶ Melanotic naevi
  - ▶ Malignant melanoma
- Treatment**
- ▶ No treatment is needed for gingival and oral lesions.
  - ▶ Systemic treatment must be left to the specialist.



**FIGURE 328**

Addison's disease.  
Early melanotic macule on the  
interdental papilla

**FIGURE 329**

Addison's disease.  
Generalized pigmentation on the  
gingiva and the alveolar mucosa



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## Paraneoplastic Disorders

### Malignant Acanthosis Nigricans

- Definition** ▶ Malignant acanthosis nigricans is a form of acanthosis nigricans that develops in association with an internal malignancy, usually adenocarcinoma of the stomach, and affects exclusively adults.
- Etiology** ▶ It is obscure, although a cytokine-like peptide produced by the malignant tumour may affect the epidermal cells.
- Gingival Involvement** ▶ Relatively rare  
▶ Non-dental plaque-induced gingival lesions
- Other Involvement** ▶ Oral mucosa, relatively common (about 30 %–40 %)  
▶ Other mucosae (conjunctiva, anus, vagina, pharynx, oesophagus), rare  
▶ Skin, always
- Main Clinical Features** ▶ The gingival lesions present as a diffuse papillomatous overgrowth, of normal colour, that may cover the crowns of the teeth (**Fig. 330**).  
▶ The oral mucosal lesions also present as diffuse velvety papillary lesions, of normal colour, particularly on the lips, commissures, tongue, and palate.  
▶ Similar lesions may develop in other mucosae.  
▶ The skin lesions appear as brownish, asymptomatic, papillary and rough growth, usually on the axillae, the genitofemoral region and the neck (**Fig. 331**).  
▶ The mucocutaneous lesions are benign, but are important as they represent a marker for internal malignancy.
- Diagnosis** ▶ The diagnosis is usually based on clinical criteria.  
▶ Biopsy and histopathological examination.
- Differential Diagnosis** ▶ Familial acanthosis nigricans  
▶ Darier's disease  
▶ Cowden's syndrome  
▶ Multiple papillomas  
▶ Lipoid proteinosis
- Treatment** ▶ The mucocutaneous lesions may resolve when the malignancy is treated.  
▶ Dental plaque control and occasionally gingivectomy for the gingival lesions.

FIGURE 330

Malignant acanthosis nigricans.  
Generalized papillomatous  
overgrowths of the gingiva



FIGURE 331

Malignant acanthosis nigricans.  
Brownish papillary overgrowths  
on the skin



### Paraneoplastic Pemphigus

**Definition** ▶ Paraneoplastic pemphigus is a unique, rare variant in the pemphigus group usually associated with lymphoreticular malignancy.

**Etiology** ▶ Autoimmune. Autoantibodies against desmoplakins I and II are a specific marker for paraneoplastic pemphigus.

**Gingival Involvement** ▶ Relatively common  
▶ Non-dental plaque-induced gingival lesions

**Other Involvement** ▶ Oral mucosa, always  
▶ Other mucosae (conjunctiva, pharynx, oesophagus, penis, vagina), relatively common  
▶ Skin, always



**FIGURE 332**

Paraneoplastic pemphigus.  
Erythema and erosions  
on the gingiva in a patient with  
pemphigus associated with chronic  
lymphocytic leukaemia

**Main Clinical Features**

- ▶ The gingival lesions present as painful, generalized and persistent erosions identical to those seen in pemphigus vulgaris (**Fig. 332**).
- ▶ The oral lesions present as multiple, severe and treatment-resistant erosions on the lips, buccal mucosa, tongue, palate, usually extending to the oropharynx and oesophagus, causing severe sore throat. Nikolsky's sign is positive.
- ▶ Persistent bilateral conjunctival erosions and oedema are common. In some cases, ocular lesions result in shrinkage and scarring.
- ▶ The skin lesions are polymorphous and include bullae, erythematous and/or verrucous papules or plaques mimicking the "target-like" lesions seen in erythema multiforme.

**Diagnosis**

- ▶ The clinical diagnosis should be confirmed by laboratory tests.
- ▶ Biopsy and histopathological examination.
- ▶ Direct immunofluorescence.
- ▶ Indirect immunofluorescence.
- ▶ Immunoprecipitation study.

**Differential Diagnosis**

- ▶ Pemphigus vulgaris
- ▶ Bullous pemphigoid
- ▶ Cicatricial pemphigoid
- ▶ Linear IgA disease
- ▶ Erythema multiforme
- ▶ Stevens-Johnson syndrome

**Treatment**

- ▶ Systemic corticosteroid alone or in combination with immunosuppressive drugs.
- ▶ Treatment of the underlying malignancy is necessary.

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## Vitamin Deficiency

### Scurvy

- Definition** ▶ Scurvy is a rare vitamin C deficiency that is usually limited to individuals (infants and elderly edentulous) whose diets are poor in fresh fruits and vegetables.
- Etiology** ▶ Vitamin C (ascorbic acid) deficiency leading to impairment of peptidyl hydroxylation of procollagen and a reduction in collagen formation and secretion by connective tissue. This deficiency leads to capillary fragility that correlates the haemorrhagic features and poor healing of wounds.
- Gingival Involvement** ▶ Common. Dental plaque-induced gingival lesions modified by vitamin C deficiency.
- Other Involvement** ▶ Oral mucosa  
▶ Skin, hair, nails  
▶ Muscles, joints
- Main Clinical Features** ▶ The gingival lesions, which present early, are common and cause generalized swelling and redness of the interdental and marginal gingiva (**Fig. 333**). Spontaneous gingival haemorrhage and ulcers are also common. Periodontal bone loss and tooth mobility may also occur.  
▶ Oral mucosal petechiae, haemorrhages, ecchymoses and delayed wound healing are commonly seen, as well as enamel hypoplasia of developing teeth.  
▶ Haemorrhage, ecchymoses of the skin, nail beds, muscles and joints are common.  
▶ Severe to moderate anaemia is common. Melena, haematuria and orbital haemorrhages may be found.  
▶ Retrobulbar, subarachnoid, and intracerebral haemorrhages may also occur if treatment is delayed.
- Diagnosis** ▶ The diagnosis is mainly based on clinical criteria.  
▶ Platelet and plasma ascorbic acid levels are reduced.
- Differential Diagnosis** ▶ Necrotizing ulcerative gingivitis  
▶ Herpetic gingivostomatitis  
▶ Agranulocytosis  
▶ Leukaemia  
▶ Thrombocytopenic purpura
- Treatment** ▶ Ascorbic acid therapy  
▶ Plaque control



FIGURE 333

Scurvy.  
Generalized gingival swelling  
associated with haemorrhages



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## PART IV

**Cysts, Tumours, Potentially Malignant,  
Neoplasms and Fibro-Osseous Lesions**

## Cysts

### Periapical Cyst

- Definition** ▶ A cyst arising from cell rests of Malassez as a consequence of inflammatory changes
- Etiology** ▶ Cell rests proliferate in response to bacterial infection of a necrotic pulp.
- Gingival Involvement** ▶ Common
- Main Clinical Features** ▶ Most are small, asymptomatic and present as a periapical radiolucency.  
 ▶ Larger cysts may present clinically with:
  - Swelling (**Fig. 334**).
  - Pain if infected.
  - Non-vital associated tooth.
  - Rarely, pathological fracture.
- Diagnosis** ▶ Diagnosis is from history and clinical features of a non-vital tooth, plus imaging.
- Differential Diagnosis** ▶ Other odontogenic cysts
- Treatment** ▶ Enucleation plus tooth extraction or endodontics. If the cyst remains after extraction, it is termed a residual cyst.
  - Residual cysts, where one of the inflammatory cysts (usually a radicular cyst) remains after removal of the non-vital tooth from which it arose (**Fig. 335**)



FIGURE 334

Odontogenic cyst.  
Radicular cyst



FIGURE 335

Odontogenic cyst.  
Residual cyst



### Eruption Cyst

**Definition** ▶ A cyst arising from the follicle of an erupting tooth

**Etiology** ▶ Eruption cysts are considered as minor soft tissue forms of follicular cysts. They often burst spontaneously before eruption of the associated tooth and only rarely require excision if impeding normal eruption.

**Gingival Involvement** ▶ Uncommon

**Other Sites of Involvement** ▶ None

**Main Clinical Features** ▶ A bluish fluctuant swelling over an erupting tooth (**Figs. 336, 337**)

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Radiography.
- Others.

**FIGURE 336**

Odontogenic cyst.  
Eruption cyst

**FIGURE 337**

Odontogenic cyst.  
Eruption cyst



**Differential Diagnosis** ▶ Other odontogenic cysts

**Treatment** ▶ Usually none

### Gingival Cyst of Newborn

**Definition** ▶ Cyst on the alveolus of neonates

**Etiology** ▶ Probably developmental, arising from remnants of the dental lamina

**Gingival Involvement** ▶ Common

**Other Sites of Involvement** ▶ None

**Main Clinical Features** ▶ These can be small, multiple, white nodules on the alveolar ridge of newborns and young infants (**Fig. 338**).

FIGURE 338

Odontogenic cyst.  
Gingival cyst of newborn



**Diagnosis** ► The diagnosis of childhood gingival cysts is usually based upon clinical features alone.

**Differential Diagnosis** ► Other odontogenic cysts

**Treatment** ► Gingival cysts in children do not require therapy.

### Gingival Cyst of Adult

**Definition** ► Cyst on gingiva of adults

**Etiology** ► Gingival cysts of adults arise from epithelial rests in the gingivae.

**Gingival Involvement** ► Common

**Other Sites of Involvement** ► None

**Main Clinical Features** ► Gingival cysts in adulthood usually arise in middle to late life as painless swellings of the interdental or attached gingivae, often in the anterior mandible (Figs. 339, 340).

**Diagnosis** ► Diagnosis is from history and clinical features, supplemented by:

- Histopathological examination.
- Radiographic assessment is essential in adults to exclude any lesion likely to be causing bony damage, or any intraosseous pathology spreading to the gingivae.

**Differential Diagnosis** ► Other odontogenic cysts

**Treatment** ► Gingival cysts in adults should be removed to confirm diagnosis. There are no long-term complications.



**FIGURE 339**

Odontogenic cyst.  
Gingival cyst of adults

**FIGURE 340**

Odontogenic cyst.  
Gingival cyst of adults



### Palatine Papilla Cyst

**Definition** ▶ A cyst in the palatine papilla

**Etiology** ▶ These cysts are derived from the vestigial oro-nasal ducts.

**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ None

**Main Clinical Features** ▶ They may occur either within the nasopalatine canal or in the soft tissues of the palate at the opening of the canal, grow slowly, and may discharge into the mouth giving a salty taste (**Fig. 341**).

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Radiography. This cyst is often discovered during routine radiographic examination. The defect appears on radiographs as a round

FIGURE 341

Odontogenic cyst.  
Cyst of palatine papilla



or teardrop-shaped, well circumscribed radiolucency. Due to its location, it can easily be misdiagnosed as a lesion of endodontic origin. Radiological examination shows a well-defined rounded ovoid or occasionally heart-shaped defect in the anterior maxilla. Nasopalatine cysts must be distinguished from a normal large anterior palatine fossa, which may be up to 7 mm in diameter, and from radicular cysts associated with the maxillary incisors.

- ▶ Histopathology may help.

#### Differential Diagnosis

- ▶ Other odontogenic cysts
- ▶ Endodontic lesions

#### Treatment

- ▶ These cysts should be enucleated and seldom recur.

### Dentigerous Cyst

#### Definition

- ▶ An odontogenic cyst (follicular cyst) that surrounds the crown of an unerupted tooth

#### Etiology

- ▶ Follicular cysts arise by separation of the dental follicle from around the anatomical crown of an unerupted tooth. Fluid accumulates between the crown and reduced enamel epithelium of an impacted tooth from degeneration of the stellate reticulum, or an accumulation of fluid between the layers of the reduced enamel epithelium.

#### Gingival Involvement

- ▶ Rare

#### Other Sites of Involvement

- ▶ Any tooth-bearing location in the jaws

#### Main Clinical Features

- ▶ They are most common in association with mandibular third molars and maxillary canines.
- ▶ Most are small, asymptomatic and found as a radiolucency surrounding the crown of an impacted tooth.
- ▶ These cysts may grow to a large size and displace the tooth with which they are associated. Larger cysts may present clinically with swelling

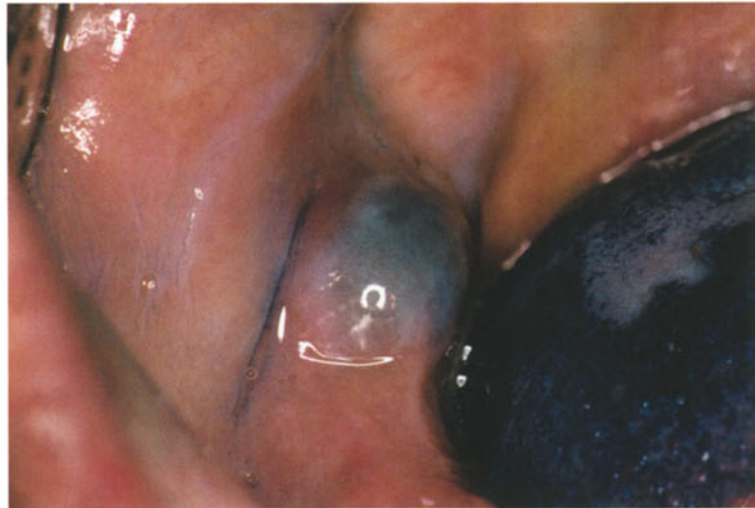
FIGURE 342

Odontogenic cyst.  
Dentigerous cyst



FIGURE 343

Odontogenic cyst.  
Dentigerous cyst



(Figs. 342, 343), pain if infected, resorption of adjacent teeth or tooth missing from arch.

- ▶ Rarely, there is a pathological fracture.

#### Diagnosis

- ▶ Diagnosis is from history and clinical features, supplemented by:
  - Radiography and histopathology; the lining of follicular cysts typically consists of flattened stratified epithelium.
- ▶ Radiographic assessment; when the follicular space exceeds 5 mm from the crown, it is likely that a cyst is present. However, odontogenic keratocysts and ameloblastomas may occasionally mimic the radiological appearance of follicular cysts.
- ▶ Aspiration may assist the diagnosis since the demonstration of keratin squames in a cyst aspirate is virtually diagnostic of a keratocyst. A protein level of less than 4.0 g/100 ml suggests a keratocyst, whereas a value over 5 g/100 ml suggests a radicular or follicular cyst, or rarely a cystic ameloblastoma.



**Differential Diagnosis** ▶ Other odontogenic cysts

**Treatment** ▶ Enucleation or marsupialization

### Keratocyst

**Definition** ▶ These cysts (primordial cyst) have generally been thought to arise from remnants of the dental lamina (cell rests of Serres) or its derivatives, but there is some suggestion that they may also arise from basal cells of the oral mucosa.

**Etiology** ▶ Idiopathic. Some cases are genetic and seen in Gorlin's syndrome.

**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ None

**Main Clinical Features** ▶ Many are seen in the mandible, especially posteriorly, and present as a uni- or multi-locular radiolucent swelling with a well-demarcated margin.  
 ▶ They involve the mandibular angle and may expand to occupy the major part of the mandibular ramus.  
 ▶ They are painless, unless they become secondarily infected.  
 ▶ Grow to a large size before giving rise to symptoms (**Fig. 344**).  
 ▶ They may resorb the cortical plates and expand into the soft tissues, may envelop unerupted teeth, giving a dentigerous appearance, or may displace teeth.

**Diagnosis** ▶ Diagnosis is from history and clinical features.  
 ▶ Histopathology. The cyst lining usually has a characteristic appearance of a regular keratinized stratified squamous epithelium, commonly five to eight cells thick. Desquamated keratin is often present within the cyst lumen and the fibrous wall is usually thin.  
 ▶ Biochemistry. Aspiration may assist the diagnosis since the demonstration of keratin squames in a cyst aspirate is virtually diagnostic of a keratocyst. A protein level of less than 4.0 g/100 ml suggests a keratocyst, whereas a value over 5 g/100 ml suggests a radicular or follicular cyst, or rarely a cystic ameloblastoma.

**Differential Diagnosis** ▶ Other odontogenic cysts  
 ▶ Nasopalatine cyst  
 ▶ Nasolabial cyst  
 ▶ Solitary bone cyst  
 ▶ Aneurysmal bone cyst  
 ▶ Stafne bone cavity

**Treatment** ▶ Small unilocular lesions should be thoroughly enucleated.  
 ▶ When the cysts are large or multiloculated, excision with a margin of surrounding bone is more appropriate.  
 ▶ Keratocysts tend to recur following removal.

FIGURE 344

Odontogenic cyst.  
Keratocyst



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The term “epulis” is applied to any lump arising from the gingiva.

Epulides are localized gingival swellings and rarely true neoplasms, metastatic carcinomas or other tumours.

### Fibrous Epulis and Denture-Induced Granuloma (Epulis Fissuratum)

- Definition** ▶ The fibrous epulis resembles a fibroepithelial polyp, but also usually has an inflammatory component.
- Etiology** ▶ Probably chronic irritation
- Gingival Involvement** ▶ Fibrous epulides are common.  
▶ Epulis fissuratum may be induced by trauma from a denture flange.
- Other Sites of Involvement** ▶ None
- Main Clinical Features** ▶ The variable inflammatory changes account for the different clinical presentations from red, shiny, and soft lumps to those which are pale, stippled and firm. Commonly, epulides are swellings which are typically firm, round, painless, pedunculated pale swellings (**Figs. 345–349**) less than 2 cm in diameter.  
▶ Arise from an anterior interdental papilla the marginal or papillary gingiva and sometimes adjacent to sites of irritation (e.g. a carious cavity).
- Diagnosis** ▶ The diagnosis is clinical, but most epulides should be removed and examined histologically.
- Differential Diagnosis** ▶ Abscesses  
▶ Exostoses  
▶ Hyperplastic gingivitis  
▶ Fibrous epulis  
▶ Cysts  
▶ Giant cell tumour  
▶ Pyogenic granuloma  
▶ Neoplasms
- Treatment** ▶ Fibrous epulides should be removed down to the periosteum, which should be curetted thoroughly.



FIGURE 345

Gingival epulis fissuratum



FIGURE 346

Gingival epulis fissuratum



FIGURE 347

Gingival fibrous lump



FIGURE 348

Gingival fibrous lump



FIGURE 349

Gingival fibrous lump



### Pyogenic Granuloma

**Definition** ▶ Pyogenic granuloma is a soft proliferative lesion that readily bleeds.

**Etiology** ▶ Possibly a reactive vascular lesion

**Gingival Involvement** ▶ Uncommon except as pregnancy epulides (lesions themselves not histologically distinguishable)

**Other Sites of Involvement** ▶ The tongue, or rarely the lip

**Main Clinical Features** ▶ Typically it is a mass that is seen on the gingival margin (Figs. 350–352) and is small (< 3 cm), red, painless, bleeds easily, ulcerates and grows rapidly.

**Diagnosis** ▶ Diagnosis is from history and clinical features supplemented by histopathology.

FIGURE 350

Gingival pyogenic granuloma



FIGURE 351

Gingival pyogenic granuloma



FIGURE 352

Gingival post-extraction granuloma





- Differential Diagnosis**
- ▶ Abscesses
  - ▶ Exostoses
  - ▶ Hyperplastic gingivitis
  - ▶ Fibrous epulis
  - ▶ Cysts
  - ▶ Giant cell tumour
  - ▶ Pyogenic granuloma
  - ▶ Neoplasms
  - ▶ Kaposi's sarcoma
  - ▶ Angiomatous proliferations
  - ▶ Chancre
- Treatment**
- ▶ Surgical excision

### Giant Cell Granuloma (Giant Cell Epulis, Peripheral Giant Cell Granuloma)

- Definition**
- ▶ This is a non-neoplastic swelling of proliferating fibroblasts in a highly vascular stroma containing many multinucleate giant cells.
- Etiology**
- ▶ May result from proliferation of giant cells persisting after resorption of deciduous teeth. The lesion probably arises because chronic irritation triggers a reactionary hyperplasia of mucoperiosteum and excessive production of granulation tissue.
- Gingival Involvement**
- ▶ Common
- Clinical Features**
- ▶ Classically, the most notable features (**Figs. 353–355**) are that it:
    - Has a deep red colour, although older lesions tend to be paler.
    - Arises interdentally.
    - Is seen adjacent to permanent teeth which have had predecessors, so is seen anterior to the permanent molars.
    - Is a benign lesion.
- Diagnosis**
- ▶ Diagnosis is from history and clinical features, supplemented by:
    - Histology, which shows many multinucleated cells distributed widely throughout the lesion or gathered into clumps.
  - ▶ Imaging.
  - ▶ Giant cell granulomas unless they are extraosseous, may also be a feature of hyperparathyroidism, and thus levels of plasma calcium, phosphate, and alkaline phosphatase should be assayed.
- Differential Diagnosis**
- ▶ Abscesses
  - ▶ Exostoses
  - ▶ Hyperplastic gingivitis
  - ▶ Fibrous epulis
  - ▶ Cysts
  - ▶ Giant cell tumour
  - ▶ Pyogenic granuloma
  - ▶ Neoplasms
- Treatment**
- ▶ Treatment depends on cause. If hyperparathyroidism, that condition must be treated. Treatment otherwise is surgical excision.
  - ▶ Histopathologically, remove local irritants (calculus). The lesion may recur.

FIGURE 353

Gingival peripheral  
giant cell granuloma



FIGURE 354

Gingival peripheral  
giant cell granuloma



FIGURE 355

Gingival peripheral  
giant cell granuloma



### Central Giant Cell Lesion

- Definition** ▶ This is an intra-bony destructive giant cell lesion.
- Etiology** ▶ The true nature of these lesions is unknown but, as they are invariably destructive, the term “reparative” giant cell granuloma would appear to be inappropriate.
- Clinical Features** ▶ Bony swelling
- Mostly in the mandible
  - Typically anteriorly
  - Only in the tooth-bearing regions of the jaws
  - Sometimes displaces or resorbs tooth roots
  - Occasionally erodes through the cortical bone where it presents as a domed, purplish submucosal swelling
  - May be symptomless or simulate a malignant neoplasm clinically and imaging
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:
- Imaging: an ill-defined area of radiolucency.
  - Histology: multinucleated giant cells irregularly distributed in a cellular stroma of plump, spindle-shaped cells, which is often highly vascular.
- Differential Diagnosis** ▶ Abscesses
- ▶ Exostoses
  - ▶ Hyperplastic gingivitis
  - ▶ Fibrous epulis
  - ▶ Cysts
  - ▶ Giant cell tumour
  - ▶ Pyogenic granuloma
  - ▶ Neoplasms
- Treatment** ▶ Curettage
- ▶ Central giant cell granulomas may recur but virtually never metastasize.

### Haemangioma

- Definition** ▶ Numerous thin-walled vessels separated by sparse bands of collagenous tissue.
- Etiology** ▶ Haemangioma is a lesion of blood vessels generally regarded as a developmental lesion (hamartoma) and usually present from birth.
- Gingival Involvement** ▶ Rare
- Other Sites of Involvement** ▶ Haemangiomas are:
- Usually isolated.
  - Occasionally a component of Sturge-Weber, Klippel-Trenaunay-Weber, Maffucci, Blue Rubber-Bleb naevus or other syndromes.
- Main Clinical Features** ▶ A haemangioma usually:
- Is deep-red (even blue-purple) in colour (**Fig. 356**).
  - Blanches on pressure.
  - Is fluctuant to palpation.



FIGURE 356

Gingival haemangioma



- ▶ It may have a lobulated or raised surface. Occasionally haemangiomas develop phlebolithiasis.

- Diagnosis**
- ▶ Diagnosis is from history and clinical features.
  - ▶ Aspiration may aid diagnosis.
  - ▶ Imaging is useful to show osseous involvement.
  - ▶ Arteriography may be indicated.

- Differential Diagnosis**
- ▶ Lymphangioma

- Treatment**
- ▶ Haemangiomas are at risk from trauma and prone to excessive bleeding if damaged (e.g. during tooth extraction).
  - ▶ Haemangiomas are left alone unless causing symptoms, when they are best treated with cryosurgery, or by laser, injection of sclerosant, ligation or embolization of feeding vessels.

### Lymphangioma

- Definition**
- ▶ Lymphangioma is of similar structure to a haemangioma, but contains lymph rather than blood.

- Etiology**
- ▶ Congenital

- Gingival Involvement**
- ▶ Rare

- Other Sites of Involvement**
- ▶ Lymphangiomas usually:
    - Are solitary
    - Affect the tongue
    - And are sometimes associated with cystic hygroma

- Main Clinical Features**
- ▶ It often has a “frogspawn” appearance (**Figs. 357, 358**).

FIGURE 357

Gingival lymphangioma



FIGURE 358

Gingival lymphangioma



- Diagnosis**
- ▶ Diagnosis is from history and clinical features.
  - ▶ Aspiration may aid diagnosis.
  - ▶ Imaging is useful to show osseous involvement.
  - ▶ Lymphangiography may be indicated.

- Differential Diagnosis**
- ▶ Haemangioma

- Treatment**
- ▶ Small lymphangiomas need no treatment but cryotherapy can be useful. Larger lesions may require excision.

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**Erythroplasia**

**Definition** ▶ Erythroplasia (erythroplakia) is an isolated red lesion.

**Etiology** ▶ Unclear

**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ Any oral site, but erythroplasia usually involves the floor of the mouth, the ventrum of the tongue, or the soft palate.

**Main Clinical Features** ▶ Erythroplasia is a red velvety lesion level with, or depressed below, the surrounding mucosa (**Figs. 359, 360**).

- ▶ Diagnosis is from history and clinical features, supplemented by:
- Histopathology; virtually all the lesions show dysplasia, carcinoma in situ, or invasive carcinoma.

**Differential Diagnosis** ▶ Other causes of red lesions, especially inflammation and candidiasis

**Treatment** ▶ Surgical excision

**FIGURE 359****Gingival erythroplakia**

FIGURE 360

Gingival erythroplakia



### Keratoses/Leukoplakia

**Definition** ▶ Leukoplakia is the term used for hyperkeratotic white mucosal lesions of unknown cause. Without specific histopathological connotations.

**Etiology** ▶ Leukoplakia and other keratoses include the following:

- Idiopathic: most cases.
- Friction.
- Tobacco: pipe smoking can cause “nicotinic stomatitis” of palate especially. Smokeless tobacco or betel-chewing can cause keratosis, usually of buccal sulcus.
- ▶ Microorganisms.
  - *Candida albicans*. Candidal leukoplakias frequently appear speckled, often affect commissures and have fairly high premalignant potential.
  - Syphilis. Syphilitic leukoplakia especially affects dorsum of tongue and has high premalignant potential.

**Gingival Involvement** ▶ Common in frictional keratosis

**Other Sites of Involvement** ▶ Any oral site

**Main Clinical Features** ▶ Most keratoses (**Figs. 361–365**) are:

- Benign.
- Seen in the buccal mucosa.
- Smooth plaques (homogeneous leukoplakias).
- ▶ Some are:
  - Warty (verrucous leukoplakia) (**Fig. 364**).
  - Mixed white and red lesions (speckled leukoplakias) (**Fig. 365**).
- ▶ Overall, 1%–3% are premalignant. Premalignant potential is higher in verrucous leukoplakias and is highest in speckled leukoplakias. Some studies show malignant transformation in over 20% of speckled leukoplakias and 30% of verrucous leukoplakias over 10 years. Thus keratoses in particular sites or of particular appearance tend to have higher premalignant potential, but epithelial dysplasia, microscopically, is a more reliable guide as may be DNA ploidy.

FIGURE 361

Gingival leukoplakia.  
Early homogeneous



FIGURE 362

Gingival leukoplakia.  
Homogeneous



FIGURE 363

Gingival leukoplakia.  
Homogeneous





FIGURE 364

Gingival leukoplakia.  
Verrucous



FIGURE 365

Gingival leukoplakia.  
Speckled



- ▶ Frictional keratosis is usually seen at sites of trauma from teeth, also along buccal occlusal line and occasionally on the gingiva beside an outstanding tooth, or on an edentulous ridge. It is homogeneous, benign and resolves when irritation is removed.
- ▶ Smoker's keratosis (stomatitis nicotina) is usually caused by pipe or heavy cigarette smoking. The palate, particularly the soft palate, is affected. Red orifices of swollen minor salivary glands of palate within widespread white lesion give striking appearance of red spots on white background. This lesion is benign in itself, but carcinoma may develop nearby.
- ▶ Other tobacco-related habits. Tobacco-chewing, snuff dipping or chewing of betel quids or ghutka may lead to keratoses that can be premalignant. Snuff dipping is associated predominantly with verrucous keratoses, which can progress to verrucous carcinoma. Tobacco-related keratoses typically resolve on stopping the habit.
- ▶ Candidal leukoplakia is seen particularly in smokers and is especially likely to form speckled leukoplakias at commissures. It may be dysplas-

tic and have higher premalignant potential than some other keratoses. Candidal leukoplakias may respond to antifungals and stopping smoking.

- ▶ Syphilitic leukoplakia is rare, but seen especially on the dorsum of tongue, and is a feature of tertiary syphilis. Premalignant potential is high.
- ▶ Proliferative verrucous leukoplakia (PVL) is a unique type of clinical oral leukoplakia, which appears related to papillomavirus and behaves in a far more aggressive fashion than other forms of leukoplakia. Its aggressiveness relates not only to a high recurrence rate, but more to a very high level of keratosis and relentless progression from a localized simple keratosis to extensive oral disease and squamous carcinomas of verrucous or conventional squamous cell type.

- Diagnosis**
- ▶ Diagnosis is from history and clinical features, supplemented by:
    - Biopsy for possible dysplasia or early malignant change.

- Differential Diagnosis**
- ▶ Materia alba
  - ▶ Burns
  - ▶ Carcinoma
  - ▶ Candidiasis (thrush: candidal leukoplakia)
  - ▶ Epstein-Barr virus (hairy leukoplakia)
  - ▶ Syphilis (syphilitic mucous patches and keratosis)
  - ▶ Lichen planus
  - ▶ Lupus erythematosus

- Treatment**
- ▶ Treat predisposing factors.
  - ▶ Surgically remove discrete lesions.
  - ▶ Consider use of retinoids.

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## Benign Neoplasms

### Fibromas

- ▶ A benign neoplasm of fibroblastic origin

### Lipoma

- Definition** ▶ A benign lesion of fatty tissue

- Etiology** ▶ Unclear

- Gingival Involvement** ▶ Rare

- Other Sites of Involvement** ▶ There are rare syndromes such as:
- Dercum's disease (multiple lipomatosis)
  - Bannayan-Riley-Ruvalcaba syndrome (macrocephaly, multiple lipomas, intestinal hamartomatous polyps, vascular malformations and pigmented macules of the penis)

- Main Clinical Features** ▶ A painless semi-fluctuant yellowish swelling (**Figs. 366, 367**).

- Diagnosis** ▶ Diagnosis is from history and clinical features.  
▶ Histopathology.

**FIGURE 366**

**Lipoma**



FIGURE 367

Lipoma



- Differential Diagnosis**
- ▶ Abscesses
  - ▶ Exostoses
  - ▶ Hyperplastic gingivitis
  - ▶ Fibrous epulis
  - ▶ Cysts
  - ▶ Giant cell tumour
  - ▶ Pyogenic granuloma
  - ▶ Neoplasms

- Treatment**
- ▶ Surgical excision

### Neurofibroma

- Definition**
- ▶ Neurofibroma represents a benign overgrowth of all elements of a peripheral nerve (axon cylinder, Schwann cells, and fibrous connective tissue), arranged in a variety of patterns.

- Etiology**
- ▶ Congenital

- Gingival Involvement**
- ▶ Rare

- Other Sites of Involvement**
- ▶ Neurofibroma is a rare lesion which typically affects the tongue. Neurofibromas may occur multiply as a feature of neurofibromatosis and may rarely undergo sarcomatous change.

- Main Clinical Features**
- ▶ Neurofibromas usually present as a swelling (**Figs. 368, 369**) and may be part of:
    - The multiple endocrine adenoma (MEA) syndrome
    - Neurofibromatosis (NF), which is now recognized as consisting of distinct variants
      - Type I (NF-I), often referred to as von Recklinghausen's disease (**Fig. 370**), which represents 90 % of all forms of neurofibromatosis. It is a disease with an incidence of 1:3000 neonates with a dominant autosomic form of transmission, but 50 % of all cases are sporadic owing to new mutations. Neurofibromas are seen mainly in NF-I.

FIGURE 368

Schwannoma



FIGURE 369

Neurofibroma



FIGURE 370

Von Recklinghausen's disease.  
Skin neurofibromas





- Type II (NF-II), a much less common disorder, which largely afflicts the central nervous system, often with bilateral acoustic schwannomas and has a more gradual onset. Neurofibromas may be seen in NF-II, but neurilemmomas and acoustic neuromas are the predominant neural tumours. Cosmetic lesions include pigmentedary changes (cafe-au-lait spots).

**Diagnosis** ▶ Diagnosis is from history and clinical features.  
 ▶ Histopathology.  
 ▶ Radiography.

**Differential Diagnosis** ▶ Abscesses  
 ▶ Exostoses  
 ▶ Hyperplastic gingivitis  
 ▶ Fibrous epulis  
 ▶ Cysts  
 ▶ Giant cell tumour  
 ▶ Pyogenic granuloma  
 ▶ Neoplasms

**Treatment** ▶ Surgical excision

### Ossifying Fibroma

**Definition** ▶ The nature of the ossifying fibroma is obscure because it has features both of a developmental anomaly and a neoplasm.

**Etiology** ▶ Idiopathic

**Gingival Involvement** ▶ Uncommon

**Other Sites of Involvement** ▶ Usually presents as a lesion arising in the jaw, sometimes only revealed coincidentally by imaging, at other times causing a swelling.

**Main Clinical Features** ▶ It presents as a painless, localized slow-growing, hard swelling of the jaw (**Figs. 371–374**).

**Diagnosis** ▶ Diagnosis is from history and clinical features.  
 ▶ Imaging: a well-defined radiolucency area with a thin sclerotic margin containing irregular opaque masses.  
 ▶ Histology: little or no distinction between ossifying fibroma and fibrous dysplasia.

**Differential Diagnosis** ▶ Abscesses  
 ▶ Exostoses  
 ▶ Hyperplastic gingivitis  
 ▶ Fibrous epulis  
 ▶ Cysts  
 ▶ Giant cell tumour  
 ▶ Pyogenic granuloma  
 ▶ Neoplasms

**Treatment** ▶ Surgical enucleation

FIGURE 371

Peripheral ossifying fibroma



FIGURE 372

Peripheral ossifying fibroma



FIGURE 373

Peripheral ossifying fibroma



FIGURE 374

Peripheral ossifying fibroma



### Chondroma

**Definition** ▶ Benign neoplasm of cartilage

**Etiology** ▶ Unclear

**Gingival Involvement** ▶ The lesion is uncommon but may present as a swelling involving the gingiva.

**Other Sites of Involvement** ▶ Usually presents as a lesion arising in the jaw, sometimes only revealed coincidentally by imaging, at other times causing a swelling.

**Main Clinical Features** ▶ This tumour is rare in the jaws. Chondromas are typically found in the elderly in the anterior maxilla, mandibular symphysis and the coronoid and condylar processes. They form slow-growing, painless masses (**Fig. 375**).  
 ▶ Multiple enchondromas are seen in the rare Ollier's syndrome but oral tumours are a rare feature of this condition.

**Diagnosis** ▶ Diagnosis is from history and clinical features.  
 ▶ Radiography.  
 ▶ Histopathology.

**Differential Diagnosis** ▶ Abscesses  
 ▶ Exostoses  
 ▶ Hyperplastic gingivitis  
 ▶ Fibrous epulis  
 ▶ Cysts  
 ▶ Giant cell tumour  
 ▶ Pyogenic granuloma  
 ▶ Neoplasms

**Treatment** ▶ If surgical excision is complete recurrence is unlikely.



FIGURE 375

Chondroma



### Salivary Adenomas

#### Pleomorphic Adenoma (Mixed Salivary Gland Tumour)

**Definition** Pleomorphic adenoma appears to originate from ductal epithelium that proliferates to contribute many duct-like spaces, sheets of epithelial cells and sometimes areas of squamous metaplasia. There are also areas reminiscent of connective tissue such as cartilage. The admixture of epithelial elements with what resembles fibrous, myxoid or cartilage tissue, leads to the name “mixed tumour”.

**Etiology** ▶ Unknown

**Gingival Involvement** ▶ Rare

**Other Sites of Involvement** ▶ Intraorally pleomorphic salivary adenomas (PSAs) are seen mainly in the palate

**Main Clinical Features** ▶ The PSA is the most common salivary gland neoplasm, usually seen in the parotid, slow-growing and benign mass. Most pleomorphic adenomas are lobulated rubbery swellings with normal overlying mucosa but a bluish appearance (**Fig. 376**). They are not fixed to deeper tissues.  
▶ Malignant change is uncommon but is suggested clinically by rapid growth, pain or fixation to deep tissues.

**Diagnosis** ▶ Diagnosis is from history and clinical features.  
▶ Imaging.  
▶ Histopathology.

**Differential Diagnosis** ▶ Other salivary tumours

**Treatment** ▶ Excision. The tumour is poorly encapsulated.

FIGURE 376

Pleomorphic adenoma



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## Malignant Neoplasms

### Oral Squamous Cell Carcinoma

**Definition** ▶ Most mouth cancer is oral squamous cell carcinoma (OSCC). Most oral cancer is cancer of the lip (International Classification of Diseases (ICD 140), tongue (ICD 141), gum (ICD 143), floor of the mouth (ICD 144) and unspecified parts of the mouth (ICD 145).

**Etiology** ▶ OSCC is seen especially in:

- Tobacco users.
- Alcohol users.
- Betel users.
- Lower socioeconomic groups.
- Ethnic minority groups.

**Gingival Involvement** ▶ Rare except in some groups, such as Japanese

**Other Sites of Involvement** ▶ Some 25% of OSCC affects the tongue – the common intra-oral site. The majority of OSCC involves the lateral border of the tongue and/or the floor of the mouth.

**Main Clinical Features** ▶ Features which suggest malignancy (**Figs. 377–380**) include:

- Erythroplasia.
- A granular appearance.
- An ulcer with fissuring or raised exophytic margins.
- Abnormal blood vessels supplying a lump.
- Induration beneath a lesion, i.e. a firm infiltration beneath the mucosa.
- Cervical lymph node enlargement.

▶ The whole mucosa should be examined as there may be widespread dysplastic mucosa (field change) or even a second primary neoplasm in the upper aerodigestive tract seen in those with OSCC over 3 years in up to 25%.

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Histopathology.

▶ Imaging is indicated to exclude local spread and second primary neoplasms.

**Differential Diagnosis** ▶ Other neoplasms

- ▶ Major aphthae
- ▶ Behçet's syndrome
- ▶ Systemic disease:
  - Infections
  - Haematological disease



FIGURE 377

Gingival squamous cell carcinoma



FIGURE 378

Gingival squamous cell carcinoma

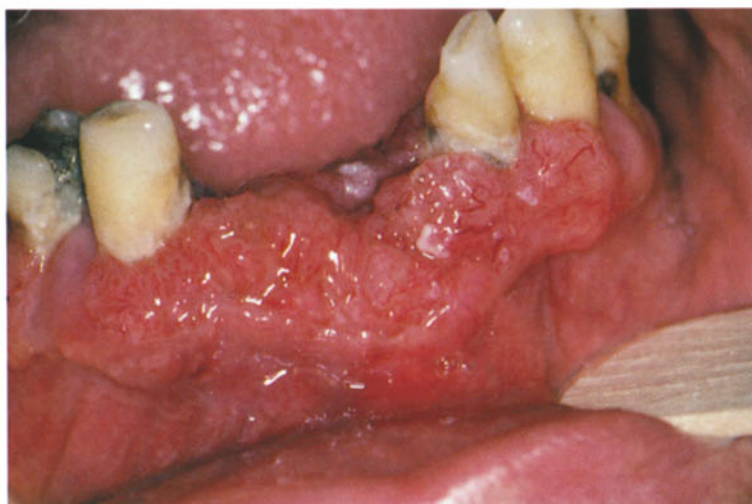


FIGURE 379

Gingival squamous cell carcinoma



FIGURE 380

Gingival squamous cell carcinoma



- Gastrointestinal disease
- Mucocutaneous disease
- Drugs
- Irradiation

**Treatment** ▶ Surgery and/or radiotherapy

### Verrucous Carcinoma

**Definition** ▶ A vegetating carcinoma that arises exophytically

**Etiology** ▶ Human papillomavirus

**Gingival Involvement** ▶ Common

**Other Sites of Involvement** ▶ Other mucosal sites (palate, buccal mucosa, tongue)

**Main Clinical Features** ▶ Nodular or warty white lesion (Figs. 381, 382).

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Biopsy.

**Differential Diagnosis** ▶ Abscesses  
 ▶ Exostoses  
 ▶ Hyperplastic gingivitis  
 ▶ Fibrous epulis  
 ▶ Cysts  
 ▶ Giant cell tumour  
 ▶ Pyogenic granuloma  
 ▶ Neoplasms

**Treatment** ▶ Surgery

FIGURE 381

Gingival verrucous carcinoma

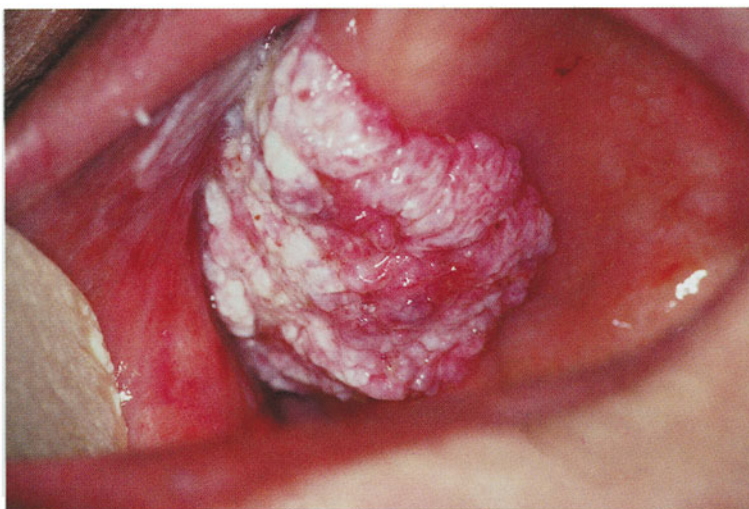


FIGURE 382

Gingival verrucous carcinoma



### Salivary Carcinomas

#### Mucoepidermoid Carcinoma

- ▶ Mucoepidermoid carcinoma accounts for up to 10% of salivary gland tumours and is a slow-growing tumour of low-grade malignancy or is benign (**Fig. 383**).

#### Acinic Cell Carcinoma

- ▶ Acinic cell tumours are very rare tumours that are usually benign, although all grades of malignancy have been reported.



FIGURE 383

Mucoepidermoid carcinoma

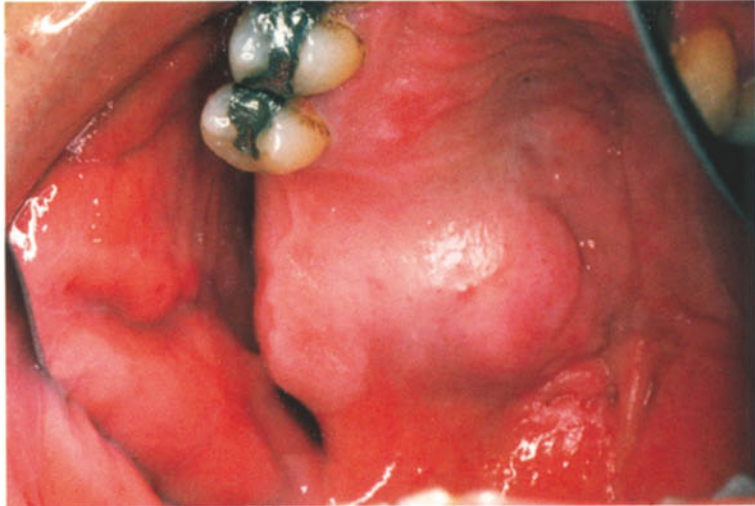
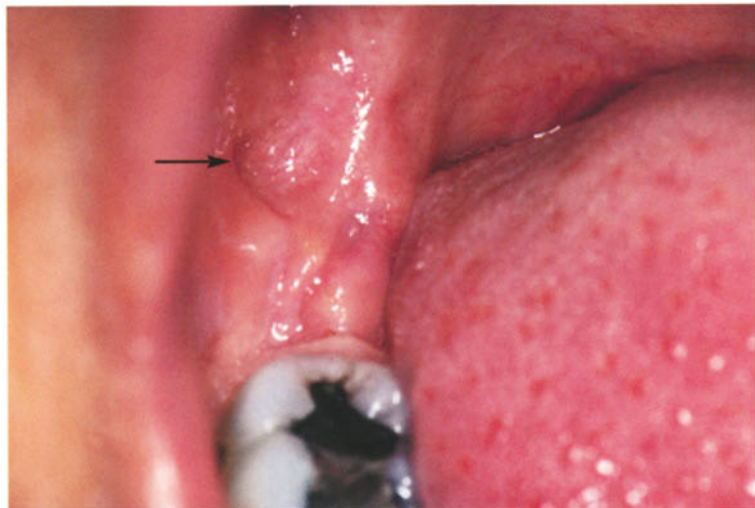


FIGURE 384

Adenocystic carcinoma



### Adenoid Cystic Carcinoma (Cylindroma)

- ▶ This is a slow-growing malignant tumour which has a tendency to infiltrate, spread perineurally, and metastasize (Fig. 384).

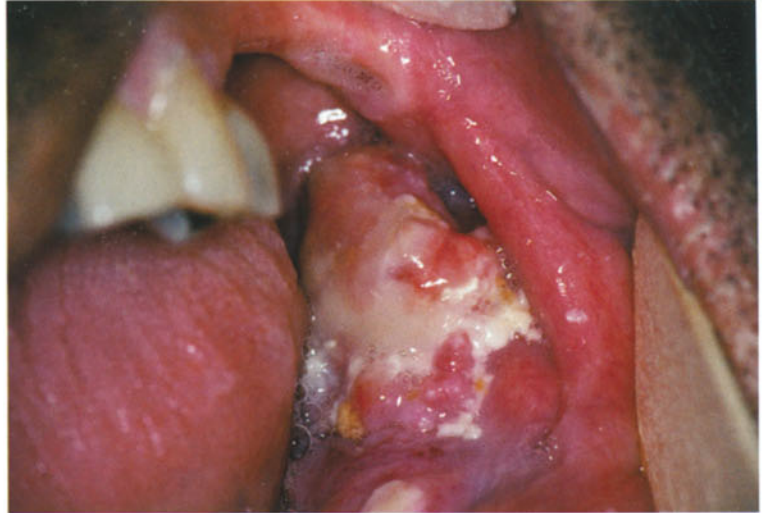
### Osteosarcoma

**Definition** ▶ Malignant neoplasm arising from osteoblasts

- Etiology** ▶ Idiopathic, although there is an increased incidence in patients who:
- Have retinoblastoma.
  - Have a history of irradiation for other lesions (about 3 % of patients with osteosarcomas).
  - Are over 50, which probably represents the small but significant malignant change in patients with pre-existing Paget's disease.

FIGURE 385

Osteosarcoma



**Gingival Involvement** ▶ Uncommon but may present as a swelling involving the gingiva

**Other Sites of Involvement** ▶ Usually presents as a lesion arising in the jaw, sometimes only revealed coincidentally by imaging, at other times causing a swelling

**Main Clinical Features** ▶ Rapidly-growing bony swelling (**Fig. 385**).

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Imaging.
- Histopathology.

**Differential Diagnosis** ▶

- ▶ Abscesses
- ▶ Exostoses
- ▶ Hyperplastic gingivitis
- ▶ Fibrous epulis
- ▶ Cysts
- ▶ Giant cell tumour
- ▶ Pyogenic granuloma
- ▶ Neoplasms

**Treatment** ▶ Resection and chemotherapy

### Ewing's Sarcoma

**Definition** ▶ A primary malignant neoplasm of undifferentiated mesenchymal cells of bone of uncertain origin

**Etiology** ▶ Unclear

**Gingival Involvement** ▶ Uncommon but may present as a swelling involving the gingiva

**Other Sites of Involvement** ▶ Usually presents as a lesion arising in the jaw, sometimes only revealed coincidentally by imaging, at other times causing a swelling

FIGURE 386

Ewing's sarcoma  
on the lower gingiva



- Main Clinical Features**
- ▶ Affects mainly children and young adults, particularly males. Rare in the jaws.
  - ▶ Tumour is extremely aggressive, expands rapidly (**Fig. 386**) and invades the soft tissues, and metastasizes early via both lymphatics and blood stream.

- Diagnosis**
- ▶ Diagnosis is from history and clinical features, supplemented by:
    - Imaging: it has either a sunray appearance or an onion-peel appearance due to deposition of new bone in layers.
    - Histopathology.

- Differential Diagnosis**
- ▶ Abscesses
  - ▶ Exostoses
  - ▶ Hyperplastic gingivitis
  - ▶ Fibrous epulis
  - ▶ Cysts
  - ▶ Giant cell tumour
  - ▶ Pyogenic granuloma
  - ▶ Neoplasms

- Treatment**
- ▶ Combination of chemotherapy and radiotherapy, has led to a 5-year survival rate of over 50%.

### Leiomyosarcoma

- Etiology**
- ▶ Idiopathic

- Gingival Involvement**
- ▶ Rare

- Other Sites of Involvement**
- ▶ Oral mucosa
  - ▶ Other sites

- Main Clinical Features**
- ▶ Non-specific swelling (**Fig. 387**).



FIGURE 387

Leiomyosarcoma on the gingiva



**Diagnosis** ▶ Diagnosis is from history and clinical features.  
 ▶ Biopsy

**Differential Diagnosis** ▶ Abscesses  
 ▶ Exostoses  
 ▶ Hyperplastic gingivitis  
 ▶ Fibrous epulis  
 ▶ Cysts  
 ▶ Giant cell tumour  
 ▶ Pyogenic granuloma  
 ▶ Neoplasms

**Treatment** ▶ Surgery

### Chondrosarcoma

**Definition** ▶ Malignant neoplasm of cartilage

**Etiology** ▶ Although isolated cases arise from chondromas, most appear de novo.

**Gingival Involvement** ▶ Uncommon but may present as a swelling involving the gingiva

**Other Sites of Involvement** ▶ Usually presents as a lesion arising in the jaw, sometimes only revealed coincidentally by imaging, at other times causing a swelling.

**Main Clinical Features** ▶ Many grow rapidly and produce extensive local destruction (**Fig. 388**).

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:  
 • Histopathology.  
 • Radiography.

**Differential Diagnosis** ▶ Abscesses  
 ▶ Exostoses  
 ▶ Hyperplastic gingivitis  
 ▶ Fibrous epulis

FIGURE 388

Chondrosarcoma



- ▶ Cysts
- ▶ Giant cell tumour
- ▶ Pyogenic granuloma
- ▶ Neoplasms

**Treatment** ▶ Radical surgery

### Metastatic Neoplasms

**Definition** ▶ Tumour deposits arising from lymphatic or haematogenous spread

- Etiology** ▶ Usually arise from carcinomas in the head and neck or from lymphoproliferative lesions, but can also arise from carcinomas especially in (decreasing order of frequency):
- Breast.
  - Lungs.
  - Kidney.
  - Thyroid.
  - Stomach.
  - Colon.
  - Prostate.
- ▶ Metastases can also arise from sarcomas (e.g. angiosarcoma, leiomyosarcoma, osteosarcoma).

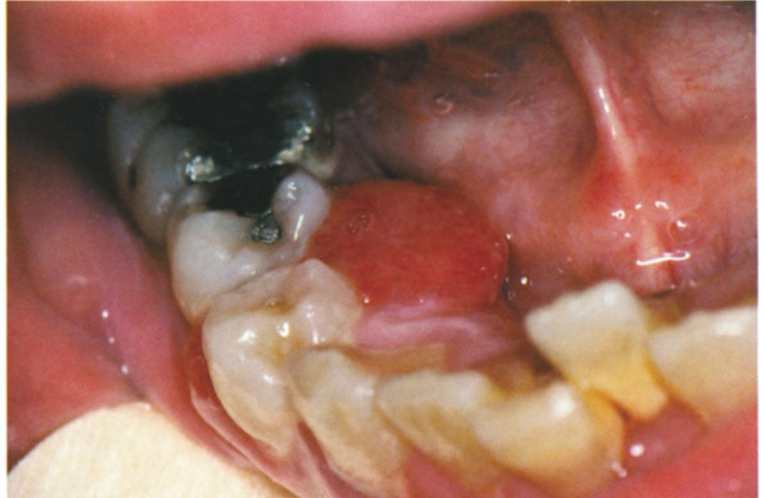
**Gingival Involvement** ▶ Uncommon but may present as a swelling involving the gingiva or periodontal destruction

**Other Sites of Involvement** ▶ Usually presents as a lesion arising in the jaw, sometimes only revealed coincidentally by imaging, at other times causing symptoms. In up to one-third of patients, the jaw lesions are the first manifestation of the tumour.

**Main Clinical Features** ▶ Metastases to the jaws mainly affect the premolar/molar region of the mandible (**Fig. 389**). Non-Hodgkin's lymphomas are frequently gingi-

FIGURE 389

Gingival metastasis  
from breast cancer



val or faucial in location. Many metastases are asymptomatic but others manifest with:

- Pain
- Paraesthesia or hypoaesthesia
- Swelling
- Tooth mobility
- Non-healing extraction sockets
- Pathological fracture
- Radiolucency or radio-opacity

**Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:

- Radiography.
- Histopathology.

**Differential Diagnosis** ▶ Infections and other causes of radiolucencies.  
 ▶ Metastases from the prostate tend to be osteoplastic and may be confused imaging with chronic forms of osteomyelitis, Paget's disease and cemental lesions.  
 ▶ Abscesses.  
 ▶ Exostoses.  
 ▶ Fibrous epulis.  
 ▶ Cysts.  
 ▶ Giant cell tumour.  
 ▶ Pyogenic granuloma.  
 ▶ Other neoplasms.

**Treatment** ▶ Treatment of the primary lesion  
 ▶ Radiotherapy, surgery or chemotherapy



FIGURE 390

Classic Kaposi's sarcoma



### Classic Kaposi's Sarcoma

- Definition** ▶ Kaposi's sarcoma (KS) is an endothelial cell multicentric malignant neoplasm characterized by increased capillary growth and prominent spindle-shaped cells with surprisingly few mitoses, extravazation of erythrocytes, and the appearance of hemosiderin.
- Etiology** ▶ Found mainly in elderly Jewish patients and Africans
- Gingival Involvement** ▶ Common
- Other Sites of Involvement** ▶ The palate, gingiva, tongue, or other oral sites (**Fig. 390**)  
 ▶ Multifocal, widespread lesions involving skin, lymph nodes and visceral organs such as the gastrointestinal tract, lung, liver and spleen
- Main Clinical Features** ▶ Early lesions of oral KS typically present as a pigmented macule.  
 ▶ Later lesions present as red, bluish, or purple nodules, sometimes ulcerated.  
 ▶ Non-discoloured oral KS is also seen.  
 ▶ KS may also present as cervical lymph node or salivary gland swellings.
- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:  
 • Histology.
- Differential Diagnosis** ▶ Racial pigmentation  
 ▶ Localized irritation, e.g. smoking  
 ▶ Amalgam tattoo  
 ▶ Naevus  
 ▶ Melanotic macule  
 ▶ Malignant melanoma  
 ▶ Epithelioid angiomatosis  
 ▶ Drugs, e.g. antimalarials  
 ▶ Addison's disease

- Treatment** ▶ Single and multiple chemotherapy agents. Laser excision, radiotherapy, cryotherapy or intralesional vinblastine or interferon-alpha may produce regression of oral KS.

## Melanoma

- Definition** ▶ Malignant neoplasm of melanocytes

- Etiology** ▶ Sunlight exposure is causal in skin melanomas, which have increased in almost epidemic fashion over the past decades, especially in older fair-skinned peoples.

- Gingival Involvement** ▶ Rare

- Other Sites of Involvement** ▶ Skin  
▶ Any oral location, particularly the palate

- Main Clinical Features** ▶ Early:
- Brown or black.
  - Initially a macule.
  - Solitary.
  - Small.
  - Asymptomatic.
  - Grow rapidly, initially spreading radially and superficially.
- ▶ Later:
- Increasingly pigmented (**Fig. 391**).
  - Nodular.
  - Deeply invasive.
  - Satellite lesions.
- ▶ Occasionally melanomas are deep-spreading nodular ab initio or are non-pigmented, multiple or large.

- Diagnosis** ▶ Diagnosis is from history and clinical features, supplemented by:
- Histopathology.
- ▶ Imaging may be helpful whenever hyperpigmentation may be caused by amalgam, graphite or a foreign body.
- ▶ Photographs may be useful for future comparison.

- Differential Diagnosis** ▶ Racial pigmentation
- ▶ Localized irritation, e.g. smoking
  - ▶ Amalgam tattoo
  - ▶ Naevus
  - ▶ Melanotic macule
  - ▶ Malignant melanoma
  - ▶ Kaposi's sarcoma
  - ▶ Drugs, e.g. antimalarials
  - ▶ Addison's disease

- Treatment** ▶ Melanomas less than about 0.75 mm thick are superficial, rarely metastasize, and can be excised in their entirety with a good prognosis.
- ▶ Others have a poor prognosis but can be excised and some centres use chemo- and immunotherapy.

FIGURE 391

Malignant melanoma



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### Paget's Disease

**Definition** ▶ A disorder in which there is uncoordinated resorption and deposition of bone, producing larger but weaker and very vascular bones (osteitis deformans). Seen mainly in Anglo-Saxons and said to be a common condition affecting up to 5 % of those over 55 years of age in some areas.

**Etiology** ▶ Unknown: virus has been implicated, possibly measles, canine distemper or respiratory syncytial virus.

**Gingival Involvement** ▶ The lesion is uncommon but may rarely present as a swelling involving the gingiva.

**Other Sites of Involvement** ▶ Monostotic and polyostotic forms are recognized.

**Main Clinical Features** ▶ Jaw swelling (**Fig. 392**) which is:

- Typically progressive
- Sometimes with pain
- Sometimes with hypercementosis

▶ Complications may include:

- Difficult tooth extractions
- Postoperative bleeding
- Postoperative infection
- Constriction of cranial foramina leading to cranial neuropathies
- Malignant change
- Arteriovenous shunting, leading to cardiac failure

**FIGURE 392**

Paget's disease



- Diagnosis**
- ▶ Diagnosis is from history and clinical features.
  - ▶ Histopathology (osteolytic stage).
  - ▶ Imaging.
  - ▶ Scintigraphy useful for revealing early and polyostotic lesions.
  - ▶ Serum alkaline phosphatase: raised.
  - ▶ Urinary hydroxyproline: raised.
- Differential Diagnosis**
- ▶ Bone neoplasms
  - ▶ Other fibro-osseous lesions such as fibrous dysplasia
  - ▶ Other conditions with a raised alkaline phosphatase, e.g. osteomalacia, hyperparathyroidism and osteoblastic metastatic deposits (e.g. carcinoma of the prostate)
- Treatment**
- ▶ Bisphosphonates or calcitonin slow the pathological process.

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**Appendix**

## Classification of Gingival Lesions by Signs and Symptoms

TABLE 1

### Keratinizing

### Non-keratinizing

Gingival white lesions

Frictional keratosis  
 Smoker's keratosis  
 Leukoplakia  
 Leukoplakia due to sanguinaria  
 Hairy leukoplakia  
 Candidal leukoplakia  
 Chronic mucocutaneous candidiasis  
 White sponge naevus  
 Focal palmoplantar and oral mucosa  
 hyperkeratosis syndrome  
 Darier's disease  
 Lichen planus  
 Discoid lupus erythematosus  
 Psoriasis  
 Squamous cell carcinoma  
 Verrucous carcinoma  
 Others

Materia alba  
 Chemical burns  
 Pseudomembranous candidiasis  
 Syphilitic mucous patches  
 Necrotizing ulcerative gingivitis  
 Others

TABLE 2

Gingival red lesions

Dental plaque-induced gingivitis  
 Non-plaque-induced gingival lesions  
 Chronic periodontitis  
 Traumatic lesions  
 Allergic reactions  
 Drug-influenced gingivitis  
 Plasma cell gingivitis  
 Desquamative gingivitis  
 Erythematous candidiasis  
 Linear gingival erythema  
 Herpetic gingivitis  
 Granulomatous gingivitis  
 Melkersson-Rosenthal syndrome  
 Sarcoidosis  
 Crohn's disease  
 Glycogen storage disease type Ib  
 Gonococcal infection  
 Scurvy  
 Gingivitis due to radiation

Erythroplakia  
 Squamous cell carcinoma  
 Leukaemia  
 Cyclic neutropenia  
 Thrombocytopenic purpura  
 Soft tissue plasmacytoma  
 Capillary haemangioma  
 Sturge-Weber syndrome  
 Klippel-Trénaunay-Weber syndrome  
 Cystic fibrosis  
 Hereditary haemorrhagic telangiectasia  
 Sjögren's syndrome  
 Porphyria  
 Reiter's disease  
 Others

TABLE 3

## Gingival ulcers and erosions

|                                      |                                  |
|--------------------------------------|----------------------------------|
| Traumatic lesions                    | Familial neutropenia             |
| Thermal burns                        | Cyclic neutropenia               |
| Necrotizing ulcerative gingivitis    | Agranulocytosis                  |
| Necrotizing ulcerative periodontitis | Aplastic anaemia                 |
| Desquamative gingivitis              | Leukaemias                       |
| Drugs (gold and others)              | Multiple myeloma                 |
| Streptococcal gingivitis             | Non-Hodgkin's lymphoma           |
| Herpetic gingivitis                  | Erythema multiforme              |
| Aphthous ulcer                       | Toxic epidermal necrolysis       |
| Behçet's disease                     | Pemphig vulgaris                 |
| Tuberculosis                         | Cicatricial pemphigoid           |
| Syphilis                             | Bullous pemphigoid               |
| Measles                              | Linear IgA disease               |
| Chickenpox                           | Pemphigoid gestations            |
| Herpes zoster                        | Epidermolysis bullosa acquisita  |
| Gonococcal infection                 | Hereditary epidermolysis bullosa |
| Cytomegalovirus infection            | Dermatitis herpetiformis         |
| Histoplasmosis                       | Chronic ulcerative stomatitis    |
| Mucormycosis                         | Lichen planus                    |
| Paracoccidioidomycosis               | Lupus erythematosus              |
| Langerhans cell histiocytosis        | Mixed connective tissue disease  |
| Squamous cell carcinoma              | Graft-versus-host disease        |
| Other neoplasms                      | Crohn's disease                  |
| Pyostomatitis vegetans               |                                  |
| Others                               |                                  |

TABLE 4

## Gingival pigmented lesions

|                          |                    |
|--------------------------|--------------------|
| Physiologic pigmentation | Lentigo            |
| Bismuth deposition       | Freckles           |
| Lead deposition          | Naevi              |
| Metal tattoo             | Malignant melanoma |
| Other tattoo             | Addison disease    |
| Smoker's melanosis       | Others             |
| Pencil peel              |                    |
| Eruption cyst            |                    |

TABLE 5

## Gingival bullous lesions

|                                 |                          |
|---------------------------------|--------------------------|
| Herpetic gingivitis             | Pemphigoid gestationis   |
| Desquamative gingivitis         | Dermatitis herpetiformis |
| Herpes zoster                   | Pemphigus vulgaris       |
| Chickenpox                      | Paraneoplastic pemphigus |
| Erythema multiforme             | Lichen planus            |
| Cicatricial pemphigoid          |                          |
| Bullous pemphigoid              |                          |
| Bullous pemphigoid              |                          |
| Linear IgA disease              |                          |
| Epidermolysis bullosa acquisita |                          |



TABLE 6

Gingival lumps and swellings,  
localized

|                                 |                                  |
|---------------------------------|----------------------------------|
| Gingival abscess                | Bacillary angiomatosis           |
| Periodontal abscess             | Kaposi's sarcoma                 |
| Pericoronal abscess             | Non-Hodgkin's lymphoma           |
| Periodontal fistula             | Burkitt's lymphoma               |
| Exostoses                       | Cutaneous T-cell lymphoma        |
| Eruption cyst                   | Other neoplasms                  |
| Gingival cyst of newborn        | Granulomatous gingivitis         |
| Gingival cyst of adult          | Tuberous sclerosis               |
| Palatine papilla cyst           | Cowden's disease                 |
| Odontogenic cysts               | Gardner's syndrome               |
| Condyloma acuminatum            | Focal dermal hypoplasia syndrome |
| Verruca vulgaris                | Actinomycosis                    |
| Focal epithelial hyperplasia    | Crohn's disease                  |
| Pyogenic granuloma              | Others                           |
| Pregnancy granuloma             |                                  |
| Peripheral giant cell granuloma |                                  |
| Peripheral ossifying fibroma    |                                  |
| Traumatic fibroma               |                                  |
| Squamous cell carcinoma         |                                  |
| Verrucous carcinoma             |                                  |

TABLE 7

Gingival lumps and swellings,  
generalized

|                                                      |                                |
|------------------------------------------------------|--------------------------------|
| Multiple exostoses                                   | Sarcoidosis                    |
| Gingival enlargement due to phenytoin                | Pyostomatitis vegetans         |
| Gingival enlargement due to calcium channel blockers | Amyloidosis                    |
| Gingival enlargement due to ciclosporin              | Malignant acanthosis nigricans |
| Hereditary gingival fibromatosis                     | Familial acanthosis nigricans  |
| Laband's syndrome                                    | Leukaemia                      |
| Hurler's syndrome                                    | Wegener's granulomatosis       |
| Sturge-Weber syndrome                                | Others                         |
| Klippel-Trénaunay-Weber syndrome                     |                                |
| Pregnancy gingivitis                                 |                                |
| Soft tissue plasmacytoma                             |                                |
| Melkersson-Rosenthal syndrome                        |                                |
| Crohn's disease                                      |                                |

TABLE 8

## Local

## Systemic

|                             |                                      |                                    |
|-----------------------------|--------------------------------------|------------------------------------|
| Causes of gingival bleeding | Dental plaque-induced gingivitis     | Cyclic neutropenia                 |
|                             | Non-plaque-induced gingivitis        | Agranulocytosis                    |
|                             | Chronic periodontitis                | Thrombocytogenic purpura           |
|                             | Aggressive periodontitis             | Aplastic anaemia                   |
|                             | Necrotizing ulcerative gingivitis    | Leukaemia                          |
|                             | Necrotizing ulcerative periodontitis | Clotting defects                   |
|                             | Pregnancy gingivitis                 | Drugs/anticoagulants               |
|                             | Pregnancy granuloma                  | Sturge-Weber syndrome              |
|                             | Pyogenic granuloma                   | Klippel-Trénaunay-Weber syndrome   |
|                             | – Peripheral giant cell granuloma    | Chédiak-Higashi syndrome           |
|                             | – Scurvy                             | Desquamative gingivitis            |
|                             |                                      | Primary herpetic gingivostomatitis |
|                             |                                      | Others                             |

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